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Surgery of Kidney and Ureteral Anomalies

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М. Д. ДЖАВАД-ЗАДЕ, Э. М. ШИМКУС

ХИРУРГИЯ АНОМАЛИЙ ПОЧЕК И МОЧЕТОЧНИКОВ

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INTRODUCTION

The recent literature has been enriched by a large number of reports devoted to the clinical picture, diagnosis and, in some instances, the treatment of kidney anomalies. Most of them, however, are articles published in various journals. More than a quarter of a century has elapsed since Gimpelson's the last and actually the only monograph in the Soviet literature on kidney anomalies was published (1949). The time has come to systematize the accumulated data on renal anomalies because new opinions on their classification, diagnosis and treatment have since appeared.

This monograph is based on the many years' experience gained in the treatment of 1405 patients with kidney and upper urinary tract anomalies at the Clinic of Urology and Operative Nephrology of the Aliev Azerbaidjan State Institute of Postgraduate Medical Training and the Clinic of Urology of the Simferopol State Medical Institute. The embryogenesis, aetiology, pathogenesis, and diagnosis of malformations of the kidneys and upper urinary tract as well as the different types of nonoperative and surgical treatment are discussed. Data are provided on the four main clinical groups of kidney anomalies.

The authors used the data in the literature to confirm the dualistic theory of the embryogenesis of the kidneys and upper urinary tract.

The chapter dealing with anomalies in the structure of the kidneys presents the existing classifications of renal cysts, in-

cluding classification which should satisfy surgeons-urologists. The same chapter includes a description of the work and findings of Professor M.D. Dzhavad-zade, Corresponding Member of the USSR Academy of Medical Sciences, at the Urological Clinic of the Pirogov Second Moscow Medical Institute (1957-1963). A total of 341 patients with various renal cysts were in our care, which made it possible to demonstrate all variations in the clinical manifestations and diagnostic peculiarities of this anomaly. The monograph provides detailed descriptions of the various types of non-operative and operative treatment of patients, including modern methods of therapy at the stage of renal insufficiency with the use of haemodialysis and kidney transplantation.

The authors discuss anomalies in the shape and interrelations of kidneys on the basis of 206 of their own clinical follow-ups and the data in the literature.

Special attention has been focused on the combination of fused kidneys with other anomalies and surgical diseases of the kidneys, which are very often the object of operative intervention. The part of the book devoted to diagnosis positively appraises renal angiography and isotope techniques of investigation (renography and renal scanning). The operative treatment of diseases of fused kidneys based on modern renal and vascular surgery is described in detail.

On the strength of 162 of their own cases the authors describe various forms of anomalies in the positioning of the kidneys. Much attention is paid to the differential diagnosis of dystopic kidneys and of surgical diseases of the abdominal and thoracic organs.

The last chapter is concerned with anomalies in the number of kidneys, based on 696 cases, including 638 cases with double kidney, 18 with aplasia, 38 with hypoplasia and two cases with supernumerary kidneys. The section of the book on treating anomalies in the number of kidneys and upper urinary tract describes corrective and plastic operations to provide for the normal flow of urine from the upper urinary tract.

Particular attention is paid in each chapter to the description of diagnostic methods and their effectiveness, the specific features of the clinical course of renal malformations, and the expediency and methods of suggested non-operative and operative treatment with assessment of the remote results.

The authors will consider their work justified if the monograph proves valuable to the practicing urologist, surgeon, specialist in oncology, gynaecologist, and radiologist.

The authors will be grateful for any critical remarks and suggestions and will give them due consideration in their future work.

CHAPTER I

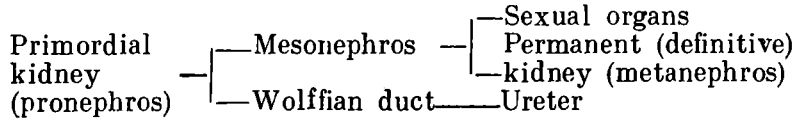
Kidney Embryogenesis

In all higher vertebrates three excretory organs are laid down in the embryonic period of development which successively replace one another.

The human kidneys undergo a very complex embryonic development. Various changes in the structure of the kidneys take place during the process of growth even after the birth of the infant.

Most scientists currently adhere to the dualistic origin of kidneys and the upper urinary tract founded by Kupffer in 1865. Later anatomo-morphological and experimental research has confirmed that the secretory apparatus of the kidney (Malpighian corpuscles, the convoluted and straight tubules, Henle's loops) is formed out of nephrogenous tissue, while the excretory apparatus (collecting tubules, calyces, pelvis and ureter) develops from the mesonephric diverticulum of the Wolffian duct.

Scheme of Kidney Embryogenesis



As the ontogenic remnant of the excretory system of lower vertebrates the primordial kidney, or head kidney, arises in humans in the third week at the stage of 9-10 somites, when the length of the embryo reaches 4.7 mm. From 7 to 10 pairs

of tubules develop at the level of 7-14 somites on both sides of the body out of the nephrotome somite. The last tubules of the primordial kidney appear at the end of the fourth week of the embryo at the stage of 23-25 somites. The cranial parts of the tubules open into the cavity of the body while the caudal parts, connecting with their neighbours, form one tube on each side. This tube is the primary excretory canal, and has been named the Wolffian duct. The tubes formed on both sides of the body elongate caudally and open into the cloaca. Glomeruli do not form in the primordial kidney and since its tubules are undeveloped they soon begin to regress gradually, beginning with those sutuated cranially, and eventually disappear. The tubules of the primordial kidney are lined with villous cells and do not fulfil an excretory function. Before the last pairs of tubules appear, only the Wolffian duct remains of the pronephros by the end of the fourth week. It is the common excretory duct of all the tubules and its distal end opens into the cloaca.

The initial elements of the mesonephros, the primary kidney, arise in the middle of the fourth week at the caudal pole of the primordial kidney. It is laid down before all the tubules of the primordial kidney regress. Thus, the mesonephros develops at the stage of 18-20 somites on the posterior wall of the second cavity of the body between the sixth cervical and third lumbar segments. Originating from the dorsal segments the segmentary limbs elongate and turn into tubules. Four tubules correspond to each segment. Developing from the posterior wall of the second cavity of the body the mesonephros raises the serous layer, thus forming a longitudinal fold known as the plica urogenitalis. Besides the mesonephros, the sexual gland and Müller's ducts are located in this fold.

In the human embryo the tubules of the mesonephros appear first as vesicles with blind ends and are located near the Wolffian duct. The vesicles gradually transform into tubules which drain into the Wolffian duct. Double-layer expansions facing the aorta are formed out of the blind ends of the tubules. These expansions were first described by Shumlyansky and Bowman and are known as the Shumlyansky-Bowman capsules. Tiny arteries branch away from the aorta toward the double-wall expansion. These arteries penetrate the expanded end of the tubules and form capillary glomeruli, called Malpighian glomeruli. The blood from these glomeruli drains along one vessel which again separates into capillaries stretching next to the tubules of the primary kidney. These arterial capillaries are then continuous with venous capillaries which drain into the general circulation. The Shumlyansky-Bowman capsule and the glomeruli together are called the Malpighian body, which in turn with the tubules

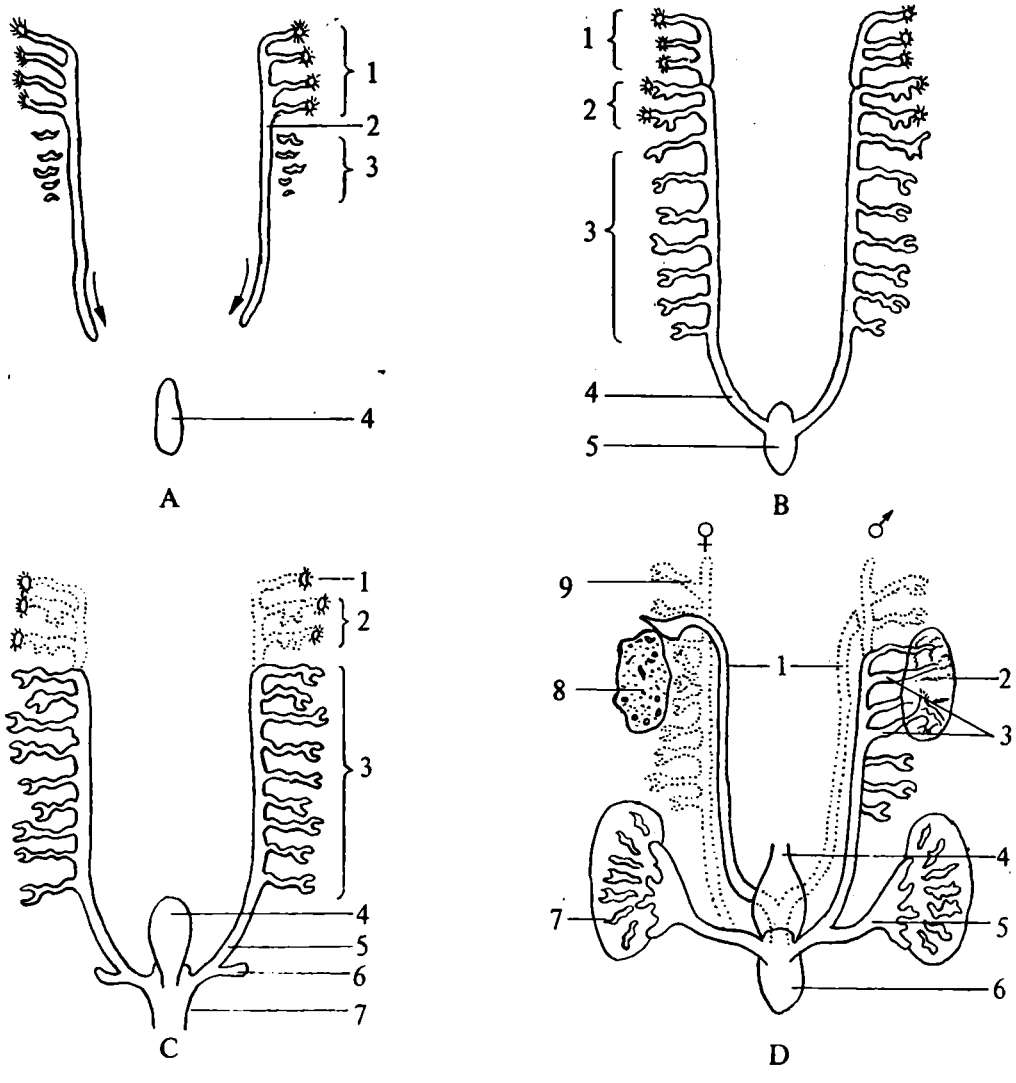


Fig. 1. Diagram of kidney embryogenesis.

A: *1*, pronephric tubules; *2*, pronephric duct; *3*, mesonephric tubules; *4*, cloaca. *B*: *1*, pronephric tubules (degenerating); *2*, mesonephric tubules with nephrostomas; *3*, mesonephric tubules without nephrostomas; *4*, mesonephric duct; *5*, cloaca. *C*: *1*, pronephric tubules; *2*, mesonephric tubules with nephrostomas; *3*, mesonephric tubules without nephrostomas; *4*, allantois; *5*, mesonephric duct; *6*, metanephric duct; *7*, cloaca. *D*: *1*, Müller's ducts; *2*, testis; *3*, mesonephric tubules; *4*, allantois; *5*, metanephric duct; *6*, cloaca; *7*, metanephric tubules; *8*, ovary; *9*, tubules degenerating into the mesonephric duct (after Patten, 1959)

of the primary kidney are called the Wolffian body, the mesonephros. The primary kidneys are located on both sides of the intestinal mesentery and this mesentery at first forms the mesentery of the primary kidney and then, continuing to ascend, forms the inguinal ligament of the primary kidneys. These mesenteries and ligaments subsequently form the ligaments of the genitals.

The tubules of the mesonephros develop in the caudal direction and by the fifth week reach its distal portion. Very soon the anterior tubules begin to degenerate and new tubules form out of the intermediate mesoderm. They are located more caudally and during development the mesonephros seems to retreat. Thus, the evolution of the mesonephros consists of successive changes and renewals of the tubules developing in the caudal and reducing in the cranial parts. The formation of the mesonephros is completed by the second month of embryonic development (Fig. 1). It then gradually atrophies, although a small part of its tubules remains in the rudimentary form. The vestige of the mesonephros in females is Gartner's duct, and in males the paradidymis.

The primary elements of the permanent kidney, the metanephros, are laid out long before the involution of the mesonephros. The permanent kidney is laid in the second month in the form of a nephrogenic cord at the level of the second and third sacral vertebrae. The permanent kidney is of dual origin. It is formed from the nephrogenic cord, the intermediate mesoderm (located caudally of the mesonephros), and from the tissues of the mesonephric duct, the Wolffian duct.

The ureter develops during five weeks of embryonic life by budding from each Wolffian duct near its entrance into the urogenital sinus. This bud, or the so-called metanephric diverticulum, grows in the cranial direction. Its cranial blind end gradually expands and eventually becomes the renal pelvis. A mass of mesodermal tissue accumulates around the pelvis, closely surrounding it and moving together with it cranially. The renal pelvis gradually penetrates this mesodermal tissue, known as metanephrogenic tissue (renal blastema), and by division gives origin to the major and minor calyces. A large number of outgrowths form on the end of each minor calyx and penetrate radially into the surrounding mass, the undifferentiated nephrogenic mesoderm. These outgrowths become hollow, forming the primary straight collecting tubules of the kidney.

By this time changes also occur in the nephrogenic tissue. Two types of tissue separate within it: the inner layer rich in nuclei, and the layer consisting of embryonic connective tissue which eventually becomes the stroma of the kidney.

Small vesicle-like groups of mesodermal cells form next to the blind end of the collecting tubule from the inner layer of nephrogenic tissue. Beginning from the second month, these vesicles elongate and turn into a tubule acquiring the S-like Shumlyansky-Bowman capsule and, together with the penetrating vascular glomeruli, form the Malpighian body of the permanent kidney.

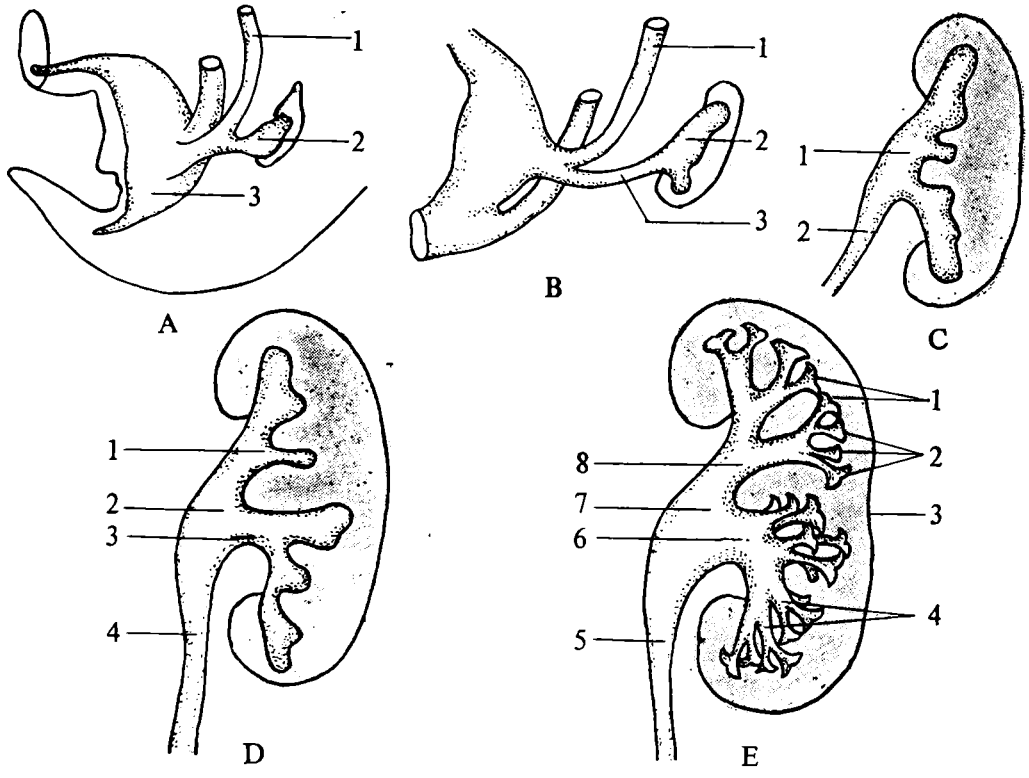


Fig. 2. Growth and differentiation of metanephric diverticulum.

A, embryo 4 mm long: *1*, mesonephric duct; *2*, metanephric diverticulum; *3*, cloaca. *B*, embryo 8 mm long: *1*, mesonephric duct; *2*, renal pelvis; 3, ureter. *C*, embryo 11 mm long: *1*, renal pelvis; *2*, ureter. *D*, embryo 13 mm long: *1*, cranial major calyx; *2*, renal pelvis; *3*, caudal major calyx; *4*, ureter. *E*, embryo 20 mm long: *1*, bundles of convoluted collecting tubules; *2*, primary straight collecting tubules; *3*, metanephrogenic tissue; *4*, developing minor calyces; *5*, ureter; *6*, caudal major calyx; *7*, transverse pelvis; *8*, cranial major calyx (after Patten, 1959)

The other end of the tube of metanephrogenic tissue is greatly elongated and forms a convoluted tubule of the first order, Henle's loop, and a convoluted tubule of the second order. The latter eventually connects with the collecting straight tubule next to which it was formed. The Malpighian body with the convoluted tubules and Henle's loop constitutes the structural and functional unit of the permanent kidney, the nephron (Fig. 2).

The differentiation of new generations of urinary tubules and glomeruli continues until the preceding generations take a definite form. The collecting tubules usually give 12 generations of branches. This branching out may continue after the birth of the infant. In the embryonic kidney of the last term of pregnancy one can see glomeruli in various stages of development, the youngest nearest the surface of the cortical layer and the functional glomeruli near the calyces. Although the permanent kidney begins functioning only toward the end of embryonic life, it how-

ever plays a very small role in metabolism. About 12 to 14 generations of tubules have formed by the time of birth and, despite the atrophy of numerous tubules of the first generations, there are about a million urinary tubules in each kidney. Several additional generations may be formed in the first few months after birth, but the increase in the kidney's dimensions in the postnatal period is evidently connected with the growth of the tubules, and not with their further increase in number.

The blood is supplied to the permanent kidney not by the aorta (as is the case with the mesonephros) but by the renal artery. This artery branches into interlobar and interlobular arteries as well as into tiny arterial branches and ends within the Shumlyansky-Bowman capsule at the distal end of the excretory tubule. The tiny arterial branch in the capsule separates into capillaries, exits from the capsule as a draining vessel and again divides into capillaries to bind closely with the remaining portion of the tubule (Patten, 1959). The venous blood is drained along the system of developing cardinal and subcardinal veins.

The secretory apparatus of the kidney (glomeruli, convoluted tubules of the first order, Henle's loop and convoluted tubules of the second order) develops from the metanephrogenic blastema, and the excretory apparatus (collecting tubules, minor and major calyces, the renal pelvis and ureter) from the vestiges of the Wolffian duct. The collecting tubules and nephrons corresponding to them form the Malpighian pyramid. The cortical substance covering the bases of the Malpighian pyramids at first has a lobular surface. Lobation of the kidney is an inherent feature of the lower vertebrates and some mammals, whereas in a five-month infant these lobes merge and by the time the child is a year old there are only traces of them left. Neither the intra- nor extrauterine development of the kidney proceeds at the same rate in all individuals. Sometimes newborn and even premature infants have kidneys which are not lobular in structure. On the other hand, there are cases where the lobation of kidneys is preserved in adults (Valker, 1938).

The permanent kidneys are located in the form of paired structures on the posterolateral walls of the body below the promontorium. At this point the hilus is in the middle of the organ and the calyces are turned medially. The permanent kidneys gradually increase in size and beginning with the seventh week migrate upward along the folds formed by the umbilical arteries.

Besides the merging of the metanephrogenic blastema (the secretory apparatus) and the excretory system, there are another three peculiarities in the development of the kidneys. These include the divergence of two metanephrogenic blastemas which

are situated practically next to each other, the upward migration of each permanent kidney and their rotation about a vertical axis. A major role here is played by the rotation of the organs, but the kidneys themselves move very little. Part of the visible migration is associated with the rapid growth of the caudal part of the body. The curvature of the embryo and the growth of the lumbosacral muscles are obviously conducive to the upward migration of the kidneys.

Before they migrate upward, the kidneys are supplied with blood by the embryonic vessels located caudally to the nephrogenic blastema. Atrophy or regression of the embryonic vessels already occurs in eight to nine weeks and the kidneys are then supplied with blood by vessels branching off from the aorta (renal artery) while the veins drain into the vena cava inferior. It is presumed that the preservation of embryonic vessels is the main obstacle to the kidney's upward migration.

By the ninth week the kidneys pass the narrow part of the coelomic cavity and again take up positions at the posterolateral walls of the body over the arterial bifurcation. During migration the kidneys rotate 90 degrees so that their convex margins, which had been directed dorsally at first, face laterally while the hilus is directed medially. The rotation or reverse rotation of the kidney usually stops when the migration of the kidney comes to an end. In their continuing movement upwards the kidneys are at the level of the second and third lumbar vertebrae by the end of the third month. The age dependence of the position of the kidneys is manifested in their gradual descent after birth. At birth they are at maximum height, and by the time the child is about two years of age they reach the position that is natural for adults. Both kidneys in a newborn are relatively frequently found at the same level (Valker, 1938).

This brief description gives an insight into the complexities of the embryogenesis of kidneys. It should be noted that the complexity of embryogenesis and the multistage development of the kidneys and upper urinary tract, as well as their intimate connections with the genitals, can be disturbed at any stage. This will be conducive to the development of various anomalies in the system. According to Vourinen (1961), for instance, anomalies so arising account for 3.5 per cent of cases.

It is not absolutely clear as yet what causes the various developmental anomalies of the kidneys and the upper urinary tract. The results of anatomical, clinical and experimental studies, however, suggest that a great variety of factors are responsible. These include infections, developmental defects in the mother, diseases involving elevation of body temperature, endometric diseases, long-term emotional suffering, diabetes mellitus, phys-

ical damage to the foetus, the effect of X-ray and atomic radiation, malnutrition, the effect of certain hormones, antibiotics, sulphanilamides and other drugs, lack of vitamins, amino acids and oxygen, various embryonic defects of the ovum and spermatozoa, etc. (Harring and Levis, 1961).

The effect of one or more of these factors during the development of the kidney inhibits or retards the growth of the organ, preventing its normal development. The influence of unfavourable factors at any particular time is also obviously conducive to similar anomalies. Bridge (1960), for example, described horseshoe kidneys in identical twins.

Some congenital diseases of the urinary system are due to damage suffered by the parents' chromosomes or the early zygote, gene mutations, changes in the chromosome number and structural aberrations.

Gene mutations include the dominant transmission of adult type polycystic kidneys; the risk of the birth of a child with polycystic kidneys in such a case could be as high as 50 per cent. In the case of polycystic kidneys of the infantile type (recessive transmission) the risk of such a repetition is no higher than 25 per cent (Savchenko et al., 1972).

Gotlieb (1926) and Higgins (1952) pointed to the role of the hereditary factor in the development of polycystic kidneys. The most fundamental work in this regard is that by Dalgaard (1957), who analysed many of his own cases of polycystic kidneys among the members of one family over the span of several generations. It proved that out of the 1949 members of the families who had been thoroughly examined 160 (10.7 per cent) had polycystic kidneys. It was also confirmed by our observations. Polycystic kidneys were registered far more often in these cases than in everyday clinical practice.

Besides polycystic kidneys, double kidneys (dominant transmission) may also be inherited. There are reports to this effect in the literature (Mikhelson, 1930; Vvedensky, 1935; Polushkina, 1939; Sethsmith and Park, 1959; Deweerd and Feeney, 1967). Whitoker and Danks (1966) studied the families of 39 patients with double kidneys. They were then found in 16 of the 123 persons examined (13 per cent). Double kidneys were encountered more often in relatives than ordinarily in clinical practice.

An increase or decrease in the chromosome number promotes the development of horseshoe kidney, double kidney, aplasia, hypoplasia, and dystopia of the kidney. Renal anomalies are encountered particularly often in the Bartholin-Patau syndrome (trisomy of chromosome 13 or trisomy 13). In this case various sexual anomalies often occur as well. In Edwards' syndrome (trisomy of chromosome 18 or trisomy 18) the urogenital system

is affected in 36 per cent of cases, in half of which a horseshoe kidney develops, in 20 per cent double kidneys, and less often aplasia, dystopia of the kidneys, megaloureter, hydronephrosis and a cyst.

Structural changes in the chromosomes are also attended by various developmental anomalies of the kidneys. Thus, hypoplasia of the kidneys was registered in Down's disease (Savchenko et al., 1972).

The following clinical classification of anomalies of the kidneys and upper urinary tract was accepted by the Second All-Union Congress of Urologists in 1978.

I. Anomalies of renal vessels

A. Anomalies of renal arteries
   1. Anomalies in number
      (a) supernumerary renal artery
      (b) duplicated arterial blood supply to the kidney
      (c) multiple renal arteries
   2. Anomalies in position (dystopia of renal artery)
      (a) low origin in the aorta (lumbar dystopia)
      (b) origin in the common iliac artery (iliac dystopia)
      (c) origin in the internal iliac artery (pelvic dystopia)
   3. Anomalies in shape and structure
      (a) genuflex renal artery
      (b) aneurysm of the renal artery
      (c) fibromuscular stenosis of the renal artery
      (d) arteriovenous fistulas

B. Anomalies of renal veins
   1. Anomalies in number
      (a) supernumerary renal vein
      (b) multiple renal veins
   2. Anomalies in shape and position
      (a) circular left renal vein
      (b) retro-aortal left renal vein
      (c) extracaval drainage of left renal vein

C. Anomalies in the interrelationship of the renal vascular bundle and other vessels
   1. Arterial aortomesenteric 'forceps'
   2. Compression of the left renal vein by gonadal arteries

II. Kidney anomalies

A. Anomalies in number
   1. Aplasia
   2. Double kidney
   3. Supernumerary kidney

B. Anomalies in size
   1. Hypoplasia

C. Anomalies in position
   1. Thoracic dystopia
   2. Lumbar dystopia
   3. Iliac dystopia
   4. Pelvic dystopia
   5. Crossed dystopia

D. Anomalies in interrelationship (fusion)
   (a) symmetrical: horseshoe, cake kidney
   (b) asymmetrical: L- and S-shaped kidney

E. Anomalies in structure
   1. Kidney dysplasia
      (a) rudimentary kidney
      (b) dwarfed kidney
   2. Cystic kidney disease
      (a) multicystic kidney
      (b) polycystic kidney disease
      (c) simple renal cysts (solitary, multilocular, dermoid)
   3. Sponge kidney

III. Anomalies of the pyelo-calyceal system
   1. Anomalies in number
      (a) pelvic aplasia
      (b) pelvic duplication, triplication, etc.
   2. Anomalies in structure
      (a) polymegacalyx
      (b) diverticulum of calyx
      (c) cyst of pelvis

IV. Ureteral anomalies
   1. Anomalies in number
      (a) aplasia
      (b) duplication, triplication, etc. (complete or incomplete)
   2. Anomalies in position
      (a) retrocaval ureter
      (b) retroiliac ureter
      (c) ectopia of ureteral orifice
   3. Anomalies in shape
      (a) corkscrew ureter
      (b) circular ureter
   4. Anomalies in structure
      (a) hypoplasia
      (b) neuromuscular dysplasia (achalasia, megaloureter, hydro-ureteronephrosis)
      (c) valves of ureter
      (d) diverticulum of ureter
      (e) ureterocele

V. Combined pyelo-ureteral anomalies

CHAPTER II

Cystic Diseases of the Kidneys

(ANOMALIES IN THE STRUCTURE OF THE KIDNEYS)

The kidneys undergo very complex embryonic development. Various changes occur in the structure of the kidneys even after birth, during the infant's growth. All this creates the possibility for deviations in the structure of kidneys and explains why there are so many renal anomalies encountered both in children and adults. Among renal anomalies a large percentage are accounted for by various cystic diseases of the kidneys. Quite often retentional, simple inflammatory cysts with multiple localizations are mistaken for polycystic kidneys. Descriptions of cases often come under the titles of polycystic kidneys, multilocular cyst of the kidney, multicystic kidney, whereas actually the problem is quite a different cystic renal anomaly. This may be explained by the complexity of the embryogenesis of the kidney and its excretory system, which hampers the correct interpretation of various forms of cystic anomalies. At the same time incorrect diagnosis of a true cystic lesion of the kidneys is often the cause of unjustified operative interventions. This is why there is such an urgent need to systematize the data on the cystic diseases of kidneys in children and adults, to clarify their pathogenesis, to make the proper conclusions essential for their diagnosis, for the recognition of the clinical picture, and for treatment. This is particularly important in establishing unilateral and bilateral cystic affection of the kidneys, because the correct identification and differentiation of unilateral affection from bi-

lateral cystic disease of the kidneys both in adults and children is of great practical significance in the choice of the method of treatment.

CLASSIFICATION

The complex and multistage development of the kidneys in the embryonal period often ends in various developmental malformations. The most complex task lies in interpreting the anomalies in the structure of the kidneys, the task being viewed by different specialists in various ways. This is why the classification of cystic renal formations proposed by different specialists can hardly satisfy all practitioners.

Although the identification of cystic structures of the kidneys has been considerably improved due to the advancement of X-ray and radioisotope diagnostic methods, these and other diagnostic techniques cannot reveal the entire pathogenesis of these anomalies and so cannot always determine the tactics to be used in treating patients with all types of cysts.

Bearing in mind that the cysts in most cases result from the disturbance or arrest of kidney embryogenesis at a certain stage and level, the approach to them from this viewpoint is fully justified.

There are currently many classifications of renal cysts, but we shall dwell here only on those that merit attention to some degree.

The first extensive clinical-experimental classification devoted to studying the pathogenesis of simple renal cysts was developed by Hepler (1930). Based on the clinical observations of seven patients suffering from solitary cyst of the kidney and experimental research the author advanced a new theory on the aetiology and pathogenesis of simple cysts.

Braasch (1940) cited a classification of the clinical forms of renal cysts; although some forms of the cystic diseases of the kidneys (multicystic kidney, sponge kidney, etc.) were not included, the classification contained great detail for its time. Later renal cysts were classified according to the forms of cystic changes, with different names given for the same forms.

Nolan (1945) classifies all cystic diseases of the kidneys as (1) simple cysts and (2) polycystic diseases of the kidneys.

Generalizing the statistical data on 50 cases of cystic diseases of the kidneys in newborn children, Potter classified them in four categories:

(1) classical polycystic kidneys with glomerular and tubular cysts;

(2) polycystic kidneys with cysts only of glomerular origin;

(3) polycystic kidneys with a corresponding reaction of connective tissue, with the development of intrarenal cartilage; (4) cysts with dilatation of the collecting tubules of the kidney.

It is evident that this classification was compiled on the basis of the histological picture of cystic changes in the kidneys.

The negative feature of this classification is the fact that Potter does not distinguish solitary, pelvic and peripelvic cysts of the kidneys encountered in children, nor does he give the names of each category.

In describing a case of a sponge kidney in a newborn infant, Andrieu offers a more correct classification of the disease occurring in children. The author claims that there is a link between sponge kidney in adults and in children; in children the disease is encountered in three forms.

In the first form the cysts are spread over all the parenchyma. The infant with such kidneys is non-viable. Microscopy reveals a very small cortex with several glomeruli and poorly differentiated convoluted tubules. The author calls this form the infantile sponge kidney.

In the second form the cortex and medulla are preserved and clearly outlined. The medulla contains many small cysts. There are only a few cysts in the cortex. A child with such a kidney may survive. Such a cystic sponge kidney is usually encountered in infants.

In the third form the cysts are usually situated only in the medulla. The cortex, although smaller than a normal one, is larger than in other forms and free of cysts. Such an anomaly is compatible with life.

It is necessary to note that the histological picture described by Andrieu in the first form greatly resembles the histological picture of a multicystic kidney. As a matter of fact, the same incorrect interpretation of a multicystic kidney occurs in the works of many researchers.

In 1959 Parkkulainen and his co-authors proposed a classification in which the cystic structures of kidneys are divided into three main groups: (1) focal cystic dysplasia, comprising five different designations within the one type; (2) multilocular cystic dysplasia comprising four different designations; (3) multicystic dysplasia comprising 12 different designations. This classification, however, fails to reveal the pathogenesis of cystic diseases.

Based on disorders in kidney embryogenesis, Osathanondh and Potter's classification provides considerable interest. Depending on the time of intrauterine development and on the component of the bud disturbance, the authors distinguish four types of cystic lesions.

Type I. Hyperplasia of the interstitial segments of the branch-

ing tubules. In this type the cysts are formed as a result of hyperplasia of the interstitium of the collecting tubules, as in the infantile type of polycystic kidneys.

Type II. In this type there is insufficient formation of nephrons because the ampullary activity of the tubules is suppressed. The initial element of the nephrons is the metanephric blastema. This means that normal differentiation of the ureteral bud does not occur. As a result the number of normally functioning nephrons diminishes, while the collecting tubules not connected with them develop cystic expansions. Multicystic kidney and renal dysplasia belong to this group of renal cysts.

Type III. Besides the defect in the metanephrogenic blastema, there is also abnormal development of the interstitial and ampullary segments of the tubules. In such a combined disturbance of kidney embryogenesis, multiple abnormal areas appear together with normally formed nephrons and collecting tubules. As a result multiple cysts form both in the cortex and the medulla. Polycystic kidneys in adults, sponge kidney, and medullary cystic disease serve as an example.

Type IV. In this group the authors included cystic diseases of the kidneys due to the reverse pressure of urine on all elements of the nephron as a result of obstructive diseases of the lower urinary tract.

This classification clarifies the pathogenesis of cystic diseases of the kidneys. It cannot satisfy clinicians, however, because although they might be interconnected different congenital forms of renal cysts differ in their clinical manifestations.

Other classifications have been proposed by Royer et al. (1963); Spense and Singleton (1972) and others. Ignatova's (1973) classification takes into account only the hereditary cystic diseases of the kidneys in children.

Pytel (1975) made a critical analysis of all the classifications of cystic lesions of the kidneys and proposed a more extensive classification. He included in it cysts resulting from renal dysplasia, polycystic kidneys in different age groups, medullary cystic lesions, simple solitary cysts, renal cysts in hereditary syndromes, calyceal and pelvic, parapelvic cysts, acquired cysts of the kidneys as well as parasitic cysts. Although such a classification encompasses all the cystic lesions of the kidneys, it is rather bulky.

With due consideration for kidney embryogenesis, clinical manifestations, and the various courses of development of cysts in adults and children, and also the fact that certain forms are acquired, we suggest the following classification. Our experience of many years shows that this classification is most acceptable for surgeonrs-uologists.

Cystic lesions of kidneys in adults:

I. Simple cysts: 1. Congenital: solitary, pelvic, peripelvic, dermoid. 2. Acquired: in pyelonephritis, in nephrosclerosis, in tumour, in tuberculosis, in hydrocalycosis.
II. Multilocular cyst.
III. Multicystic kidney.
IV. Sponge kidney.
V. Polycystic kidney disease.

Cystic lesions of kidneys in children:

I. Unilateral cystic lesions: 1. Multilocular cyst. 2. Unilateral multicystic kidney. 3. Solitary cyst. 4. Pelvic cyst. 5. Peripelvic cyst.
II. Bilateral cystic lesions: 1. Bilateral multicystic kidney. 2. Sponge kidney. 3. Polycystic kidney disease similar to polycystic disease in adults.

SIMPLE RENAL CYSTS

Simple cysts are divided into congenital (primary) and acquired (secondary) cysts.

Congenital primary cysts include the following: solitary cyst, pelvic cyst, peripelvic cysts, dermoid cyst.

SOLITARY CYST

This solitary cystic structure is either round or oval in shape. In most cases it bulges from the surface of the kidney and may be localized in various parts of the organ (Fig. 3, *A*, *B*, *C*). The contents of the cyst are usually serous, rarely haemorrhagic because of bleeding into the cavity of the cyst. The solitary cyst is the most commonly encountered among renal cysts. Its size

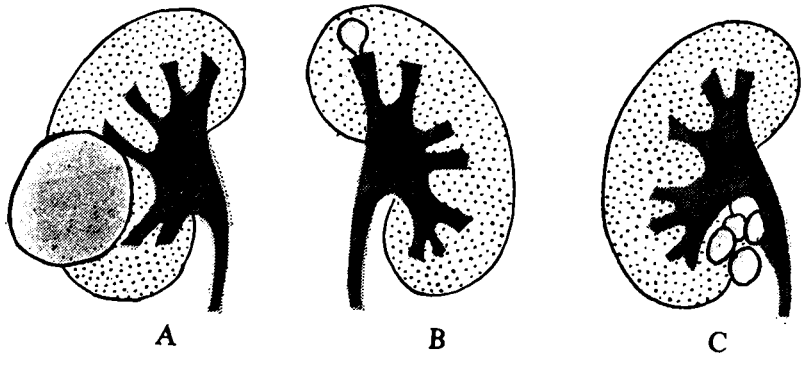


Fig. 3. Congenital renal cysts.
A, solitary cyst; *B*, pelvic cyst; *C*, peripelvic cysts

is usually no more than 8-10 cm in diameter. Very large cysts have been described, however, containing more than 10 litres of fluid.

Surgeon urologists often treat this disease surgically. But the majority of cases have not been recorded and so the value of the literature is limited.

The solitary cyst occurs about just as frequently among males as among females. Thus, of the 46 patients whom we operated on for solitary cyst of the kidney 27 were males and 19 females.

Aetiology and pathogenesis. The first serious clinical and experimental study of simple cysts of the kidney was conducted by Hepler (1930). The author advanced a new theory on the aetiology and pathogenesis of simple cysts. Experiments were performed on rabbits and dogs whose renal papilla was either ligated or destroyed; in some animals this was combined with the ligation of one of the branches of the renal artery. In the latter cases the author obtained a solitary cyst of the kidney, very similar to that occurring in man. When the artery was not ligated, the destruction or ligation of the papilla resulted only in the dilatation of the tubules. Proceeding from these experimental data, Hepler considered that a solitary cyst will develop only when circulation is disturbed in the renal parenchyma in a limited area where there is some obstacle preventing the drainage of urine along the collecting tubules.

This theory was later advocated by Gibson (1954).

Most researchers share the opinion that solitary renal cysts are retentional and develop in conditions unfavourable for the drainage of urine along the collecting tubules, caused by obstruction of the tubule and active glomerular secretion above the obstacle. The impaired urine drainage along the tubules may be caused by local inflammation of a group of tubules with peritubular sclerosis and deformity of the tubule. This hypothesis has been verified by numerous clinical and anatomo-histological studies, in which several small retentional cysts are often found on the surface of the kidney in pyelonephritis or nephrosclerosis. In bilateral and multiple localization of cysts such kidneys may be mistaken for the polycystic disease of the kidneys. The fact that retentional cysts are registered at an age older than 40 confirms the possible involvement in their formation of various arteriosclerotic changes in the renal parenchyma.

This hypothesis, however, cannot explain the development of the large, so-called solitary cysts, in whose origin a major role is possibly played by disorders in the embryogenesis of different nephrons, a result of improper connection of the excretory and secretory portions of one or more nephrons.

According to some authors, in addition to epithelial cysts,

there are lymphatic cysts both in the cortical and the medullary zones of the kidney. In their opinion, the dilatation of lymphatic vessels is due to the absence of a connection between the lymphatic system of the cortex and medulla.

Thus, solitary cysts may be congenital or acquired in origin. According to our studies, however, the cysts in most cases are of a congenital character. This was confirmed by the results of pathohistological examinations of the cystic walls: atrophy due to compression but with no signs of inflammation was found, while the inner surface of the cysts was lined with cuboidal epithelium. With regard to localization, solitary cysts are most frequently found in the lower pole and more rarely in the upper and middle portions of the kidney. Out of 46 patients with solitary cysts of the kidney, 34 had cysts in the lower pole of the kidney, seven in the middle portion and five in the upper pole.

The progressive growth of the cyst gradually leads to atrophy of the renal parenchyma. With the occurrence of infection, interstitial nephritis develops and even suppuration of the cyst. Large renal cysts also disturb renal haemodynamics, which in turn may bring about arterial hypertension; an important role in this is played by concomitant infection and the development of unilateral interstitial nephritis.

The clinical course of a solitary cyst of the kidney is extremely variable. The most characteristic signs of the disease are a dull pain in the hypochondrium or lumbar region and a palpable tumour; total haematuria is a rare occurrence. When the disease is complicated by an inflammatory process (suppuration of the cyst) the disease takes an acute course with high body temperature, chill and the intensification of pain around the kidney.

If the disease is complicated by arterial hypertension, the latter is often of a transitional character.

The diagnosis of a solitary renal cyst is not difficult. It is based on medical history, palpation and X-ray examination, although the unilateral enlargement of the kidney, which makes it accessible for palpation, as well as the various deformities of the pelvis and calyces caused by the solitary cyst often lead to the erroneous clinical diagnosis of tumour of the kidneys. This is all the more probable when the patient has a history of total haematuria caused by disorders in haemodynamics in the fornical zone of the calyces. Defects in the area of the calyces and pelvis produced on the retrograde and excretory pyelograms in solitary cysts, however, have regular, rounded contours as a rule. Renal angiography is of great value for a differential diagnosis. The solitary cysts on the angiograms have a typical pattern: a rounded, clear, avascular area (Fig. 4).

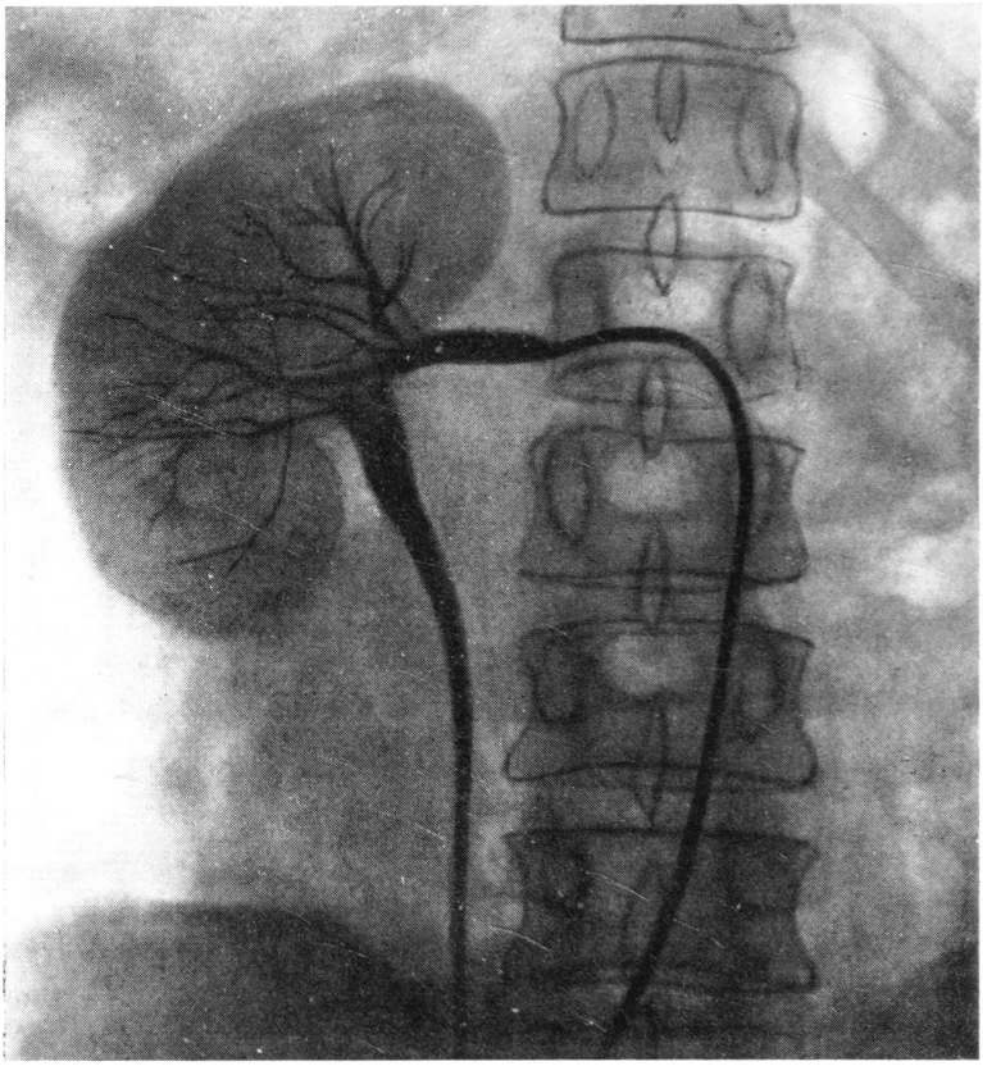


Fig. 4. Solitary cyst in lower pole of right kidney (selective renal angiography)

Patients with solitary cysts are treated by surgery. The operation consists of cutting open and emptying the cyst, with excision of the edges of the wall at the level of the normal renal parenchyma (Fig. 5). When a large cavity is left, it is packed with perirenal fat, after which the edges of the wall are sutured with interrupted catgut sutures (Fig. 6). Attempts to enucleate the cyst are neither grounded nor justified because such intervention leads to recurrent haemorrhage or the development of sites with urine retention.

In suppuration of the cyst operative intervention should be limited to the opening and drainage of the cyst. In complication

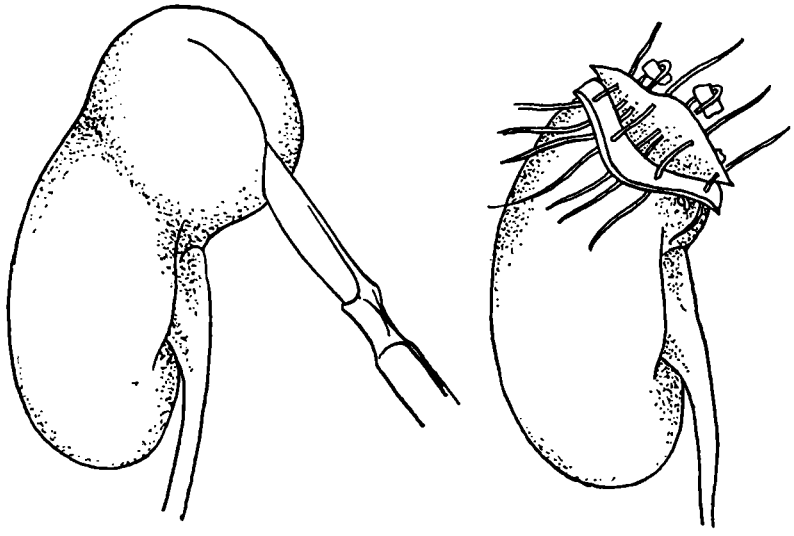


Fig. 5. Diagram of operation for resection of a solitary cyst; principles of suture application

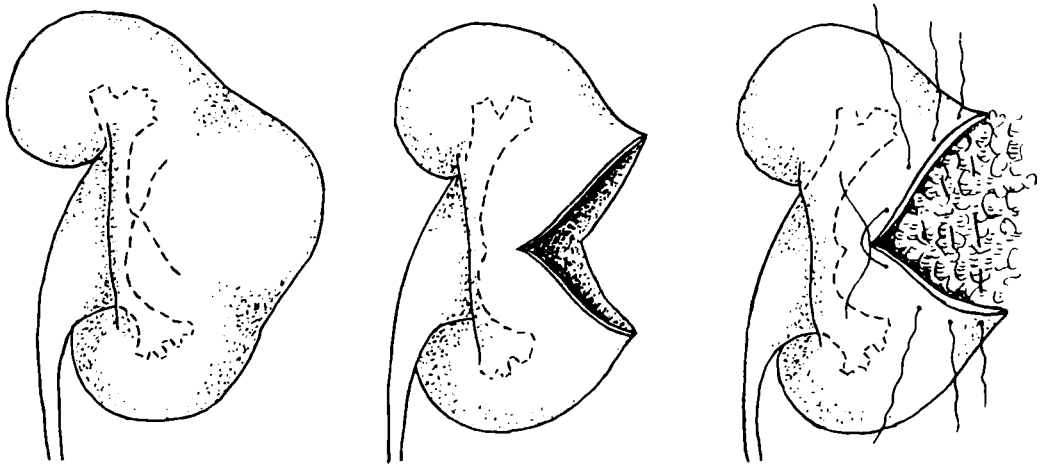


Fig. 6. Diagram of operation for removal of a solitary cyst from the mid-portion of the kidney. Tamponade of the cystic cavity with fatty tissue

of the disease by arterial hypertension, which is most often caused by interstitial nephritis and disorders of haemodynamics in this kidney, it is more expedient to perform nephrectomy if the contralateral kidney functions normally.

Postoperative prognosis and the remote results are quite favourable.

PELVIC CYST

The pelvic cyst (diverticulum of the pelvis) is encountered very rarely. According to the literature there are three types of

pelvic cysts. Heusch (1964) distinguishes the intrapelvic type, when the cyst is located on the inner surface of the pelvis and shows a tendency to grow into the pelvic cavity; and the intramural type, when the cyst is located on the outer wall of the pelvis and has a tendency for outward growth. These are 'true' pelvic cysts of congenital character. But one should also distinguish the diverticulum of the pelvis. Unlike hydrocalycosis and peripelvic cysts, the diverticulum of the pelvis is usually single and communicates with a normal calyx by means of a narrow passage (see Fig. 3*B*). By 1964 there had been only 108 cases of pelvic cyst and diverticulum of the renal pelvis reported in the literature. We encountered another two cases in our practice.

The *aetiology and pathogenesis* of solitary pelvic cysts and diverticulum of the pelvis have not been fully studied. The cause of this disease, however, lies in disorders in the embryogenesis of the kidney. Some authors share the opinion that a major role in the genesis of pelvic diverticula is played by disturbed synchronous activity of the papillo-fornical and papillo-calyceal neuromuscular apparatus. It is quite possible that circulation is disturbed in a limited area of the renal parenchyma. The epithelium lining such a cyst is the same as the epithelium of the pelvis. The pelvic diverticulum may become larger due to urine flowing into it from the pelvis. The wall of such a distended diverticulum may protrude over the surface of the kidney, giving it the appearance of a solitary renal cyst.

The permanently impaired drainage of urine from the diverticulum is conducive to the development of an inflammatory process both in the diverticulum itself and in the parenchyma of the kidney. This is why a pelvic cyst is usually attended by pyelonephritis. The clinical manifestations of the disease are associated with disturbed urine drainage from the upper urinary tract and development of an inflammatory process in the kidney. In some cases the disease is manifested by total haematuria. Haematuria may occur as a consequence of renal haemodynamic disorders but more often as a result of pyelonephritis. Cases have been described of arterial hypertension in patients suffering from pelvic cyst of the kidney. The diagnosis of this condition is extremely difficult, particularly the recognition of the pelvic cyst. Very often a correct diagnosis of the disease is impossible even during an operation. In one case described by Heusch (1964), for instance, the cyst which was palpated in the renal pelvis during operative intervention was mistaken for a tumour and a nephro-ureterectomy was performed.

The X-ray picture of a true pelvic cyst resembles a pelvic tumour because of the filling defect seen on the pyelogram.

In diverticulum of the pelvis a spherical-like cavity commu-

nicating with a normal calyx and pelvis is shown above the calyx on the pyelogram.

The treatment of this disease is surgical. A cyst arising from the wall of the pelvis is, according to Heusch, small and solitary and can be easily resected.

Nephrectomy is indicated when there is a large diverticulum of the pelvis and pyelonephritis.

The prognosis and the remote results are favourable, according to reports in the literature. Nephrectomy was performed in one of our patients; in another, the cyst was resected.

PERIPELVIC CYST

The peripelvic renal cyst is a rare disease. The first reports on this peculiar cystic kidney disease, identified at autopsy, were made about a hundred years ago. A special case of peripelvic cysts was described by Zvara (1961). Scholl (1943) and other authors described cases of peripelvic cysts revealed during operation. A total of 31 cases of typical peripelvic cysts had been reported in foreign literature before 1964. We failed to find any reports on this disease in the Soviet literature, which may be connected with lack of knowledge of this renal disease among physicians. Peripelvic cysts are multiple cystic new growths located at the hilus of the kidney, next to the pelvis and in close proximity with the vessels. Such a location and the outward appearance of peripelvic cysts may be the reason why they are often mistaken for cysts arising from the renal pelvis itself. In distinction from pelvic cysts, however, peripelvic cysts do not communicate with the pelvis (see Fig. 3*C*).

Peripelvic cysts are sharply dilated lymphatic vessels which have suffered inflammatory changes and obliteration of their lumina. This explains the deep location of the cysts in the renal sinus, their position along the length of the renal vessels and their adhesion to the renal pelvis. The fact that peripelvic cysts have an endothelial lining confirms this viewpoint. Lymphatic renal cysts are similar to other lymphatic cysts in the human body. They possess an endothelial lining. The average size of peripelvic cysts is 1 cm, although cysts as big as hen's egg have been described. The cysts usually have a multichamber structure.

Some authors contend, however, that peripelvic cysts originate from the Wolffian duct. They explain the endothelial lining by the flattening of the epithelial cells by intracystic pressure.

It is evident from the literature that a clinical diagnosis of peripelvic cyst has yet to be made. Such cysts have been found in patients in whom they were diagnosed as tumours or cysts of the kidney or polycystic kidney disease, because peripelvic cysts had

produced various changes in the pyelogram. Götzen (1960) describes four cases of a peripelvic cyst. It is interesting that in one case the cyst was attended by heightened arterial pressure. The author points out correctly that in those cases when pyelographic changes are insufficient to be confident of a diagnosis of a tumour or a renal cyst and lumbotomies are undertaken for this reason, one should bear peripelvic cysts in mind.

In 1962 a patient was operated on at our clinic with the above-mentioned cystic anomaly of the kidney. The cysts were enucleated and the lower pole resected.

Although a diagnosis of a peripelvic cyst has not yet been made prior to an operation, if multiple cysts are discovered in the area of the renal hilus during an operation, operative intervention must be organ-preserving and limited to enucleation of the cysts.

The prognosis of the disease is favourable.

DERMOID CYST

This cystic anomaly of the kidney is encountered very rarely. According to Yurikhin, only eight cases of dermoid cysts of the kidney had been described in the literature up to 1935. Dermoid cysts may consist of fat, hair, a sebaceous mass and sometimes even teeth are found.

Aetiology and pathogenesis of a dermoid renal cyst, as of dermoid cysts in general, are connected with disorders of embryogenesis.

It is impossible to establish a correct diagnosis of a dermoid renal cyst clinically. It is discovered by chance in patients who have been operated on for tumour or cyst of the kidney. A dermoid cyst of immense size growing from the upper pole of the kidney was excised in one of our patients. The cyst consisted of fatty tissue and a greenish-brown fluid. There were areas of cartilaginous consistency on the inner surface of the cyst, and hairs in the paste-like fatty mass.

After the cyst had been resected, the kidney was transposed from the iliac to the lumbar region and fixed according to Revoire. A control examination conducted three years later showed that the kidney was functioning well.

Due to its extreme rarity, this cyst is of no great practical significance in differential diagnosis.

Treatment is operative. If there is a sufficient amount of functioning renal parenchyma, operative intervention should be limited to enucleation of the cyst.

ACQUIRED CYSTS

Acquired secondary cysts are cysts occurring with pyelonephritis, nephrosclerosis, tumours and tuberculosis of the kidney. In pyelonephritis and nephrosclerosis the cysts are small in appearance (1-2 cm), filled with serous content and located on the surface of the kidneys (Fig. 7*A*). They are discovered more often during an operation on the kidney or during autopsy. These cysts are retentional because of the obliteration of several tubules as a result of inflammatory and sclerotic processes. Owing to their multiplicity and frequently bilateral location they are sometimes diagnosed erroneously as polycystic kidney disease. Such an incorrect diagnosis is often made at autopsies.

A renal cyst occurring in tumours, called a cystadenoma, is also a retention cyst. Sometimes, during a blastomatous process in the kidney, when the tumour plugs the renal tubules and a large arterial branch, a cyst may form (Fig. 7*B*). Besides, the growing tumour may compress the surrounding tissue and lead to atrophy. A retention cyst then develops where the renal parenchyma surrounds the tumour (Pytel).

In such cases one may receive erroneous impression that the tumour is growing from the wall of the cyst. Such cysts may become very large. Gibson (1954), for instance, describes a cyst in a sarcomatous renal tumour which contained 7800 ml of fluid. In most cases the contents of such cysts are haemorrhagic.

Renal cysts are often encountered in tuberculosis. However it is the multiple cysts which are of practical significance in a kidney affected by tuberculosis. The condition must be differentiated from a tuberculous polycystic kidney. An important role in the origin of cysts in tuberculosis of the kidney is played by secondary pyelonephritis, as well as by the sclerotic process in the renal

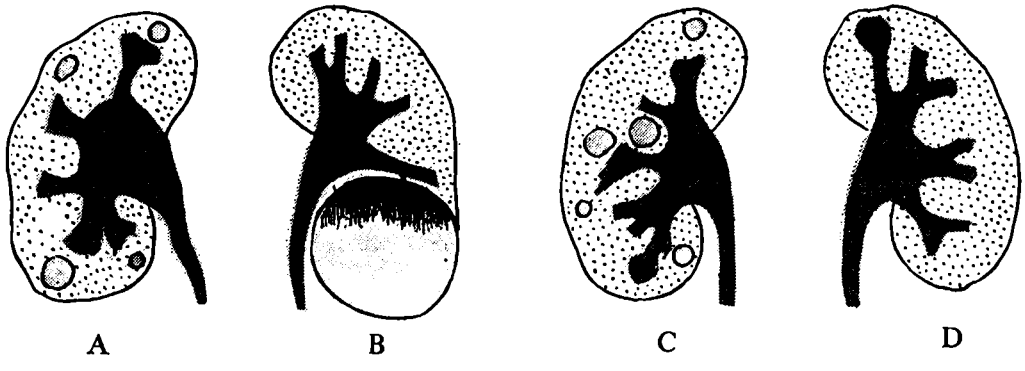


Fig. 7. Acquired secondary renal cysts.

A, cysts in pyelonephritis and nephrosclerosis; *B*, cyst in tumour of kidney; *C*, cysts in tuberculosis; *D*, hydrocalycosis

parenchyma occurring during tuberculosis: several cysts no larger than 1-2 cm and with serous contents (Fig. 7*C*) protrude above the surface of the kidney together with the caverns. In such a combination of cysts and tuberculosis of the kidneys, especially when there are fewer tuberculous caverns than cysts, and in a bilateral process, differential diagnosis with polycystic kidneys may prove to be very difficult. This is emphasized by Gasparyan (1941). Scrupulous analysis of the medical history and anatomopathological data, however, will help to avoid such a diagnostic error.

The treatment of acquired secondary cysts in tumours, tuberculosis and pyelonephritis is linked with the management of the principal disease.

HYDROCALYCOSIS (CYST OF THE CALYX)

Hydrocalycosis is retention dilatation of a calyx which makes it difficult for urine to flow out of it (Fig. 7*D*). This occurs as a result of pathological processes developing in the area of the neck of the calyx. These include infiltrates which compress and thus narrow the neck, and stones located in the calyx which may occlude the neck and cause hydrocalycosis. Another cause may be dysfunction of the muscular sphincter of the calyceal neck (van Gulik, 1959). With the accumulation of urine, the calyx dilates, forming a sphere resembling a cyst. It usually consists of turbid urine; very often the cavity of the cyst also contains a multitude of concretions. Vainberg described a case with 22 tiny concretions discovered in the cavity of the hydrocalycosis during an operation. When operating on a 22-year-old patient for hydrocalycosis of the left kidney, we found and removed 48 concretions 0.8 to 0.2 cm in size from the cavity of the cystically distended calyx. Although cases of hydrocalycosis are rarely described in the literature, the disease occurs quite frequently in practice.

It is essential to differentiate hydrocalycosis from pelvic cyst. This is not difficult as a rule, if we bear in mind that in hydrocalycosis the wall of the calyx is drastically changed, its cavity is dilated due to the narrowing or obliteration of the neck of the calyx, whereas in pelvic cyst the neck and the calyx itself are not changed.

Hydrocalycosis is clinically manifested by dull renal pain, pyuria and haematuria. The pain may have the character of renal colic at times.

When there are stones, the shadows of concretions are outlined on the scout X-ray. A filling shadow with even, rounded edges

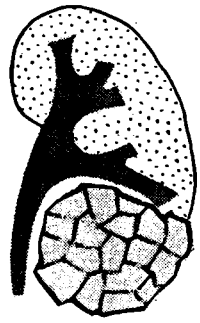


Fig. 8. Multilocular cyst (diagram)

is seen on the retrograde pyelogram. In some instances, the flow of the contrast medium through the narrowed neck into the spherical cavity is demonstrated on the pyelogram. The accumulation of the contrast medium in the spherically distended calyx may be seen on excretory urograms.

Treatment is surgical. After the kidney has been exposed, the size and location of the cyst are determined, it is opened and its contents are removed. The area of the calyceal neck is inspected closely, revealing a very narrow fissure or depression on the floor of the cavity. The neck of the calyx is widened first by the sharp technique and then by gradual bougienage so that the cavity of the calyx might communicate sufficiently with the pelvis. The edges of the wall of the cystically dilated calyx are sutured with interrupted catgut sutures. A drainage tube is introduced down to the lower pole of the kidney for three or four days.

Postoperative prognosis and the remote results are quite favourable.

MULTILOCULAR CYST

In the diagnosis of polycystic and multicystic kidney the multilocular cyst creates confusion. In describing the multilocular cyst some authors actually described a unilateral multicystic kidney, while others, in describing cases of multicystic and polycystic kidney, actually described cases of multilocular cyst. Spence (1955) and others gave a very good and detailed description of the clinical and pathological picture of the multilocular cyst of the kidney. The diagnosis of a multilocular multichamber cyst should be based on a combination of the following signs:

(1) the cyst must be unilateral and located only in one part of the kidney, with no changes in the remainder of the parenchyma;

(2) it must consist of many chambers (not communicating with one another or with the pelvis);

(3) unlike the polycystic kidney, the walls of the chambers should not contain the renal parenchyma.

The multilocular multichamber cyst develops as a result of anomalous embryonic development of the kidney. The cysto-dysplastic process is limited to one part of the kidney, whereas the remainder of the kidney develops normally. A cystically changed area resembling one large cyst (Fig. 8) develops mostly in one of the poles of the kidney. On section it proves to consist of many chambers divided by numerous septa. The cystically changed area may involve a large part of the kidney, but it is clearly demarcated from the normal parenchyma.

A multilocular cyst of the kidney occurs rarely. From 1892 to 1966 only 22 cases have been described in the literature, one of which was a case of a multilocular cyst in a 22-month-old child. Nephrectomy was performed on the patient with a favourable outcome. We have encountered multilocular cysts in three patients. Thus, only 25 cases of a multilocular cyst of the kidney have been described in the world literature to date.

We now give a description of one of our cases so as to throw light on certain questions of the clinical course, diagnosis and differential diagnosis of this cystic anomaly of the kidneys.

52-year-old woman (case history No. 5632) was admitted to the clinic on January 28, 1958 with the diagnosis: polycystic kidney disease, tumour of the left kidney. Quite by chance the patient had discovered a tumour-like structure 18 months previously in the left hypochondrium. The physician diagnosed it as a polycystic kidney disease. In the 18 months the palpable tumour in the left hypochondrium grew and began troubling the patient.

The general condition of the patient was satisfactory. No abnormalities were found in the thoracic and abdominal organs.

A hard, uneven mobile structure 20×16 cm in size was palpated in the left hypochondrium. The symptom of renal ballottement was positive. The right kidney was not palpable. Urine and blood tests, as well as Zimnitsky's test, showed normal values. Blood urea was 40.64 mg/100 ml.

Chromocystoscopy: the mucosa of the urinary bladder was normal. Indigo carmine injected intravenously was discharged on the fifth minute from both orifices. A small pelvis and drastically elongated and considerably distended calyces were demonstrated on the left retrograde ureteropyelogram. The ureter was displaced medially (Fig. 9). The excretory urogram showed good functioning of the right kidney with a normal pyelo-calyceal system. On the left the function was sharply weakened and there were changes resembling those on a retrograde ureteropyelogram.

The results of the general clinical and X-ray examination were insufficient to confirm the diagnosis of polycystic kidney disease but at the same time a malignant tumour was suspected because the process was unilateral. Renal angiography was performed by means of a translumbar puncture of the aorta to make a more exact diagnosis. Angiograms showed good filling of the abdominal aorta at the level of the whole length of the first lumbar vertebra, the unchanged right renal artery and its branches, and drastically distorted artery of the left kidney. The blood to this kidney is supplied by two arteries, one of which, a long thin trunk, supplies the lower pole, dividing, in turn,

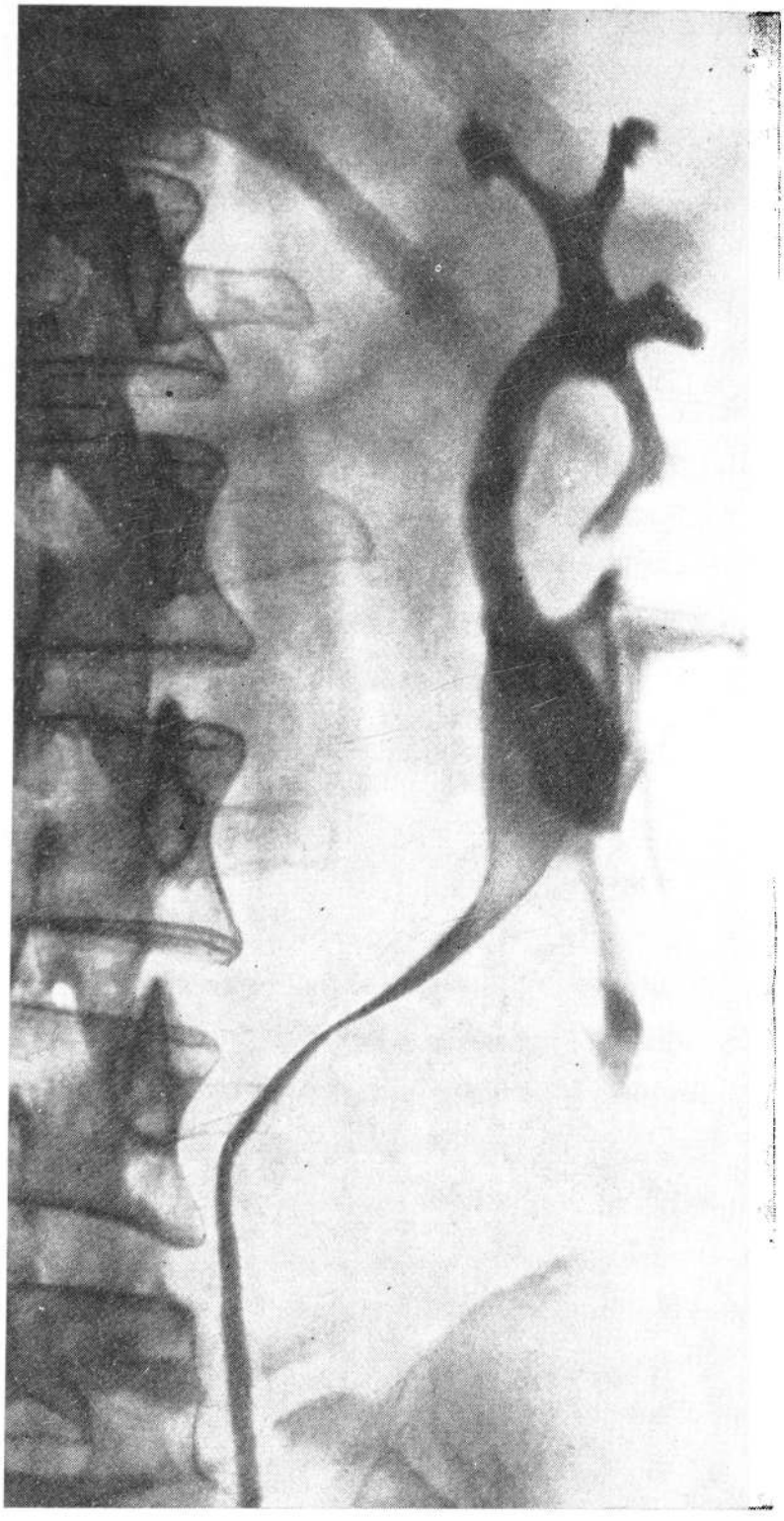


Fig. 9. Multilocular cyst in lower pole of left kidney (retrograde pyelogram)

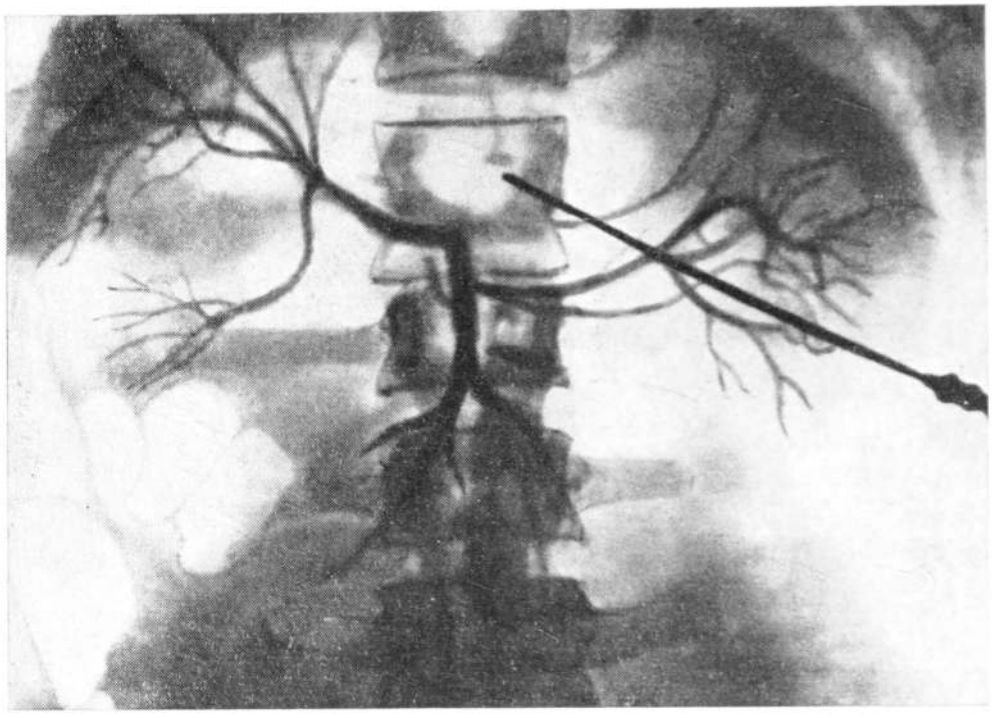


Fig. 10. Renal arteriogram (arterial phase)

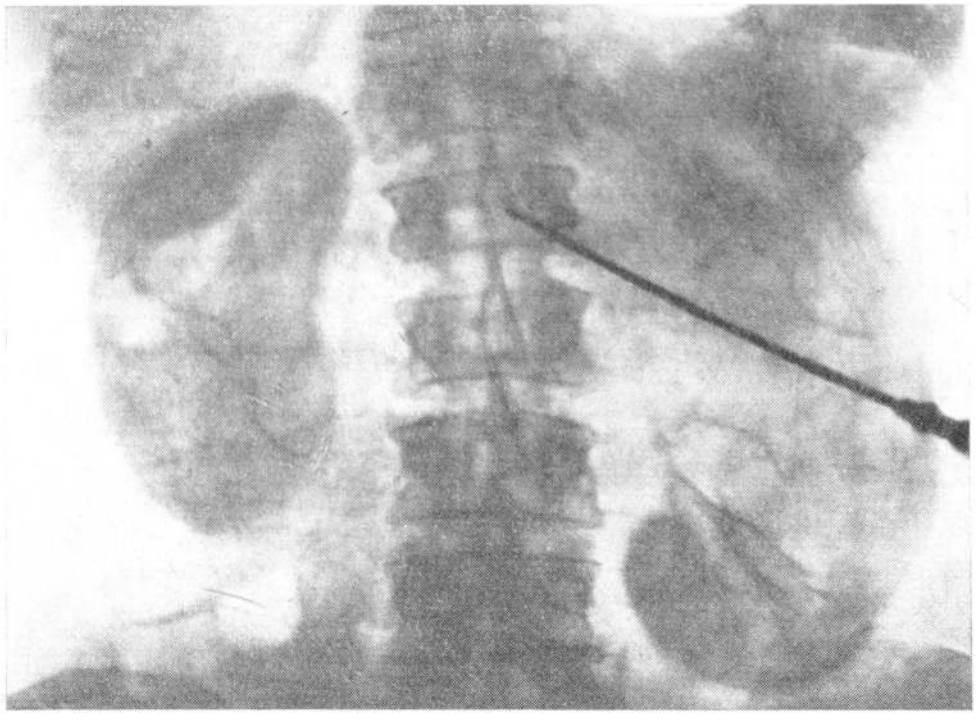


Fig. 11. Renal arteriogram (parenchymal phase)

into two thin branches between which there are avascular areas. A small amount of the contrast medium was noted in the para-aortic space (Fig. 10). The shadow of the normal right kidney was clearly outlined on the nephrogram. There was no shadow of the upper two thirds of the left kidney on the nephrogram, and only a barely discernible shadow of the lower pole (Fig. 11).

On the basis of the results of the examination, the diagnosis of tumour and polycystic kidney disease was ruled out and cystic affection of the upper two thirds of the left kidney was established. During the operation, a multilocular cyst of the left kidney with atrophic changes in the remaining third of the kidney was revealed. Nephrectomy. Recovery.

As one can see from this case and the data published in the literature on cases of multilocular cyst of the kidney, treatment of this cystic anomaly consists of nephrectomy. However, if a sufficient amount of functioning renal parenchyma remains uninvolved in the lesion, and with functional insufficiency or the absence of the contralateral kidney, surgical intervention should be limited to excision of the cyst.

Prognosis is quite favourable.

MULTICYSTIC KIDNEY

The multicystic kidney is a congenital anomaly in which the renal parenchyma is fully replaced by cysts of irregular shape and various size. If bilateral, such an anomaly of the kidneys is usually incompatible with life. Therefore bilateral multicystic kidney is encountered in stillborn and newborn infants who died immediately after birth.

Problems of the diagnosis and treatment of unilateral multicystic kidney and its differentiation from polycystic kidney merit attention. Although both diseases are developmental anomalies of embryogenesis, the consequences of abnormal development after the birth of the child are manifested differently.

The aetiopathogenesis of the multicystic kidney can be explained more correctly from the viewpoint of the dualistic theory of kidney development. According to this theory, as has already been stated, in the process of embryogenesis the excretory part (ureter, pelvis, calyces and straight collecting tubules) of the permanent human kidney forms from the mesonephric duct, while the secretory apparatus (glomeruli, convoluted tubules) forms from the intermediate mesoderm (metanephrogenic tissue). The normal development of the permanent kidney depends on the correct fusion of these two buds. Clinical and pathohistological studies have shown that the multicystic kidney forms as a result of incomplete fusion of both buds, or when there is no bud of the excretory apparatus and only the secretory apparatus of the permanent kidney develops. The developing metanephrogenic tis-

sue produces urine which is not discharged, however, and so accumulates; in this way cysts of various sizes are formed. This was confirmed by Parkkulainen and Visakorpi (1959), who examined the contents of cysts in four children operated on for multicystic kidney. The fluid in the cysts is a filtrate of long functioning glomeruli with some degree of reabsorption in the tubules. Toward the end of intrauterine life there is practically complete replacement of the metanephrogenic tissue by cysts. If there were buds of the ureters, pelvis, and calyces, they gradually atrophied because they did not communicate with the secretory apparatus and were inactive.

Our studies of all patients with polycystic disease of the kidneys have revealed that usually the condition is bilateral. This is also verified by the hereditary character of the disease. The unilateral cystic affection in most cases is the multicystic kidney and this disease is not inheritable.

Terminology. In the literature various names are often used for this disease: polycystic rudimentary kidney (Rosenow, 1911); embryonic cyst of the kidney (Galkin, 1936); unilateral polycystic kidney (Wakeley, 1930; Lih et al., 1952), aplasia with cystic degeneration (Tedeschi, 1952; Abeshouse, 1956); cystic rudiment of metanephros (Krause, 1935); congenital cystic hydrocalycosis (Fink et al., 1957); unilateral multilocular cysts (Dakin, 1940); simple multiple cysts (Christeson, 1954); congenital cystic disease with hydronephrosis (O'Donell and Presti, 1949); metanephrotic cystic tumour (Moore, 1957), and 'tuberous' kidney (Bibus, 1954), multicystic dysplasia of kidneys (Parkkulainen and Visakorpi, 1959), etc.

The diverse terminology for this disease is evidently explained by the rarity of this form of cystic affection of the kidney. The term unilateral multicystic kidney, however, proposed by Schwartz (1936) is the most adequate.

There are two variants of the position of the ureter of a unilateral multicystic kidney: (1) when the ureter is atrophied and ends blindly without reaching the urinary bladder; in such cases no ureteral orifices are seen in the bladder; (2) the orifice of the ureter of the multicystic kidney is situated at its usual site in the urinary bladder, but when filled with the contrast medium, the ureter ends blindly in the area of the renal pelvis (Fig. 12).

Macroscopically the multicystic kidney consists of numerous (up to 20 and more) cysts of different shape and size. They are usually filled with a clear or straw-coloured fluid. There is loose connective tissue between the cysts. Calcification of the walls of the cysts often occurs in adults: foci of bone tissue and cartilage are sometimes found (Fine and Burns, 1959). Cases of suppuration of the cysts have been described as well as tumour in multicystic

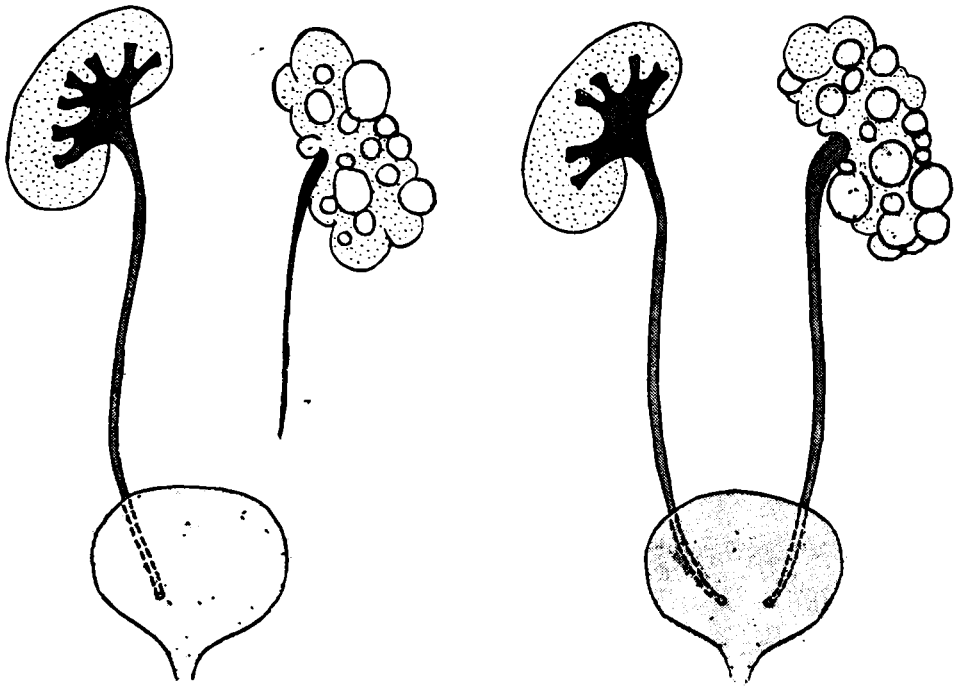


Fig. 12. Two variants of multicystic kidney

kidney (Charles, 1935). In most cases the ureter is either absent, atrophied or rudimentary. The vascular bundle is absent, there being only a few small vessels.

Microscopic study of the cysts reveals them to be lined with a single layer of cuboidal epithelium, their walls consisting of fibrous tissue, sometimes with areas of hyalinosis and foci of calcification.

In some instances there are small areas near the centre of the cystic mass containing isles of renal tubules or an undeveloped embryonic structure with clearly demarcated interstitial connective tissue. Areas of calcification or of bone and cartilaginous tissue are usually found in multicystic kidneys removed from adult patients. On rare occasions large tubules are seen lined with columnar or cuboidal epithelium surrounded by large quantities of fibrous or mesenchymal tissue. The ureter is atrophied.

The unilateral multicystic kidney, either in children or adults, occurs very rarely. Fine and Burns (1959) collected the descriptions of 48 cases from the literature and added six of their own. Fransvea and Papadia (1959) described a case of a nine-month-old boy with a unilateral multicystic kidney successfully treated by nephrectomy. In the Soviet literature Galkin (1936) and Kuzmina (1955) each reported a case of the disease. We had three patients with unilateral multicystic kidney; in each case neph-

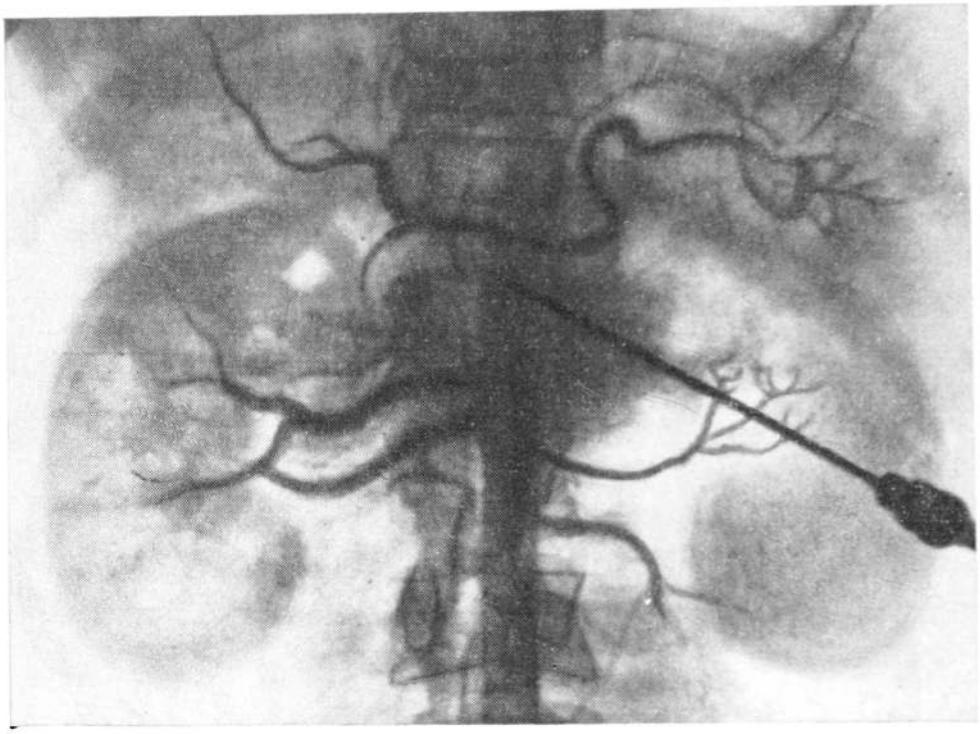


Fig. 13. Renal arteriogram (arterial phase)

rectomy was performed with good results. Problems of the diagnosis of a multicystic kidney and its differential diagnosis with polycystic kidneys are illustrated in the following case record.

E., a 31-year-old male, was admitted to the clinic on December 7, 1959, complaining of dull pain in the left lumbar region. He considered himself to have been ill since 1956, when he experienced pain here for the first time. A diagnosis of lumbosacral radiculitis was established and physiotherapy applied. Up to 0.033 pro mille protein was detected in the urine in October, 1958 on the grounds of which polycystic kidneys were suspected. The patient was sent to our clinic for a more exact diagnosis and further treatment. The relatives of the patient had not suffered from renal diseases.

The patient's general condition was satisfactory. No abnormalities were found in the organs of the thoracic and abdominal cavities. Arterial pressure was 120/65 mm Hg. A soft elastic tumour 14×8 cm in size with an uneven surface was palpated in the left hypochondrium; it was mobile. The right kidney was not palpable. The external genitalia were normally developed.

Urinalysis: specific gravity 1025; protein 0.006 pro mille; leucocytes 0-4 in field of vision; erythrocytes 2-5 in field of vision.

Blood test: haemoglobin 90 per cent; erythrocytes 4 500 000; leucocytes 9100; ESR 5 mm/hour; blood urea 27.44 mg/100 ml.

Chromocystoscopy: normal picture of the urinary bladder mucosa. The orifice of the right ureter was normal whereas that of the left ureter could not be detected, there being a small depression in its place. Indigo carmine was discharged in a good stream from the right orifice after four minutes.

No visible changes were detected on the scout urogram.

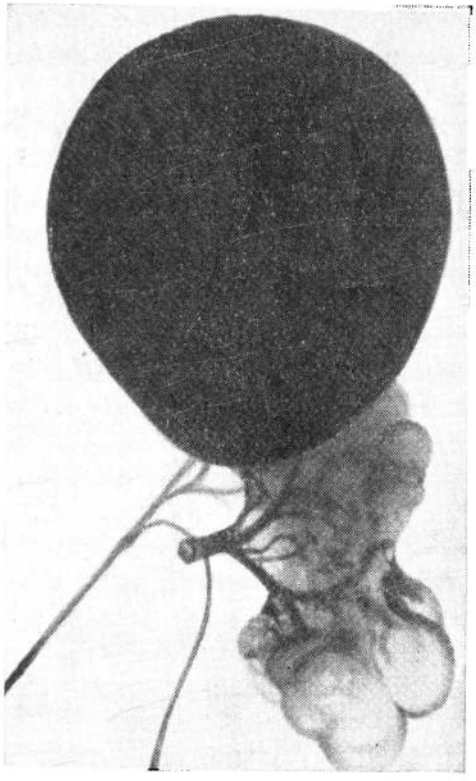


Fig. 14. Removed multicystic kidney

In 10 and 20 minutes satisfactory function of the right kidney and absence of function of the left kidney were recorded on excretory urograms. The contrast medium filled the somewhat elongated calyces, pelvis and ureter of the right kidney quite well. On the left the upper urinary tract was not outlined. Since the disease was undefined translumbar aortography was performed. Two renal arteries and the normal contours of the kidney were seen on the right of the angiogram. On the left, the renal artery was drastically thinned out and separated into three thin branches at the renal hilus. The shadow of the kidney appeared in the form of a hazy avascular structure (Fig. 13).

Diagnosis: multicystic kidney on the left. Left lumbotomy performed under intubation ether-oxygen anaesthesia. Perinephrium was poorly developed. There was a cystic tumour-like structure in place of the kidney with large cysts (4×2 cm, 3×2 cm and 1×1 cm in size) in the upper pole; the walls of the cysts were very hard. Besides small cysts, there was a hard mass in the lower pole of the structure. A band resembling a ureter stretched from the middle portion of this cystic structure. When this band was transected, however, it proved to have no lumen. A very fine artery and vein approached the upper pole; they were ligated and the kidney was removed. This was a typical picture of a multicystic kidney (Fig. 14).

Histological study showed the walls of the cyst to consist of fibrous connective tissue lined by stratified cuboidal or in places columnar epithelium. No glomeruli or tubules were seen. Between the fibrous tissue there were layers of fatty tissue and deposits of lime in some parts.

The postoperative period was uneventful and the patient was discharged on the twelfth day after the operation. Examination eight months later showed his condition to be good. He had no complaints.

A correctly made differential diagnosis between multicystic and polycystic kidney is of great diagnostic, particularly of prognostic, importance.

These two diseases, with different aetiopathogenesis and clinical course, require a correct approach in choosing the surgical intervention. Nephrectomy in multicystic kidney is indicated and has a favourable effect on the condition of patients; in polycystic kidneys it is contraindicated not only because the organ is removed, but because it is hazardous for the life of the patient since the disease is usually bilateral. The only exception is cancer and pyonephrosis of the polycystic kidney, when nephrectomy is vitally indicated.

How can one make the diagnosis and differential diagnosis of multicystic kidney and polycystic kidney?

A multicystic kidney can only be unilateral in both adults and children because such a bilateral anomaly of the kidneys is incompatible with life and occurs only in stillborn infants and in those who died immediately after birth. Polycystic kidney disease is usually a bilateral condition both in adults and children and presents a peculiar clinical picture differing from that in multicystic kidney.

Clinically the unilateral multicystic kidney is often manifested by symptoms that are not absolutely characteristic and it may be discovered by accident or at autopsy. Patients sometimes complain of spasms of pain or of dull pain in the hypochondrium or the lumbar region. These painful sensations are usually not attended by a rise in body temperature, haematuria or pyuria. A rise in arterial pressure in patients with a multicystic kidney is a rare occurrence, whereas these symptoms are encountered very frequently in patients with polycystic kidneys. Some patients have abnormalities in the intestine (pain, inflation, etc.), which are evidently to be explained by compression of the adjacent intestinal loops by the large multicystic kidney. Developmental anomalies of other organs are also observed in patients with a multicystic kidney.

In objective examination of patients with a multicystic kidney, an elastic, lobate, mobile and painless tumour-like structure may be palpated in the depth of the hypochondrium or the lateral half of the abdomen.

Cystoscopy and retrograde ureteropyelography show the absence of the ureteral orifice of a multicystic kidney or a small depression in its place. In rare cases the orifice may be found, but ureterography shows that the ureter usually ends blindly below the place where it joins the rudiment of the kidney. In adults scout urography reveals areas of calcification in the kidney where calcium is deposited in the cystic walls.

Excretory urograms usually show no function or shadow of the multicystic kidney and good function of the contralateral kidney.

Renal arteriography and presacral pneumoretroperitoneum are valuable in diagnosis, because with them it proves possible to demonstrate the shadow of a tumour-like structure at the site of the kidney. Normal angiograms and the shadow of a normal kidney are produced in renal angiography on the healthy side, but not on the affected side. It is sometimes possible to detect small vessels in the area of the renal artery on renal angiograms and a weak shadow of the multicystic kidney as was the case in one of our patients. These angiographic changes, as a rule, occur only on one side, while renal angiography in patients with polycystic kidneys demonstrates the characteristic changes in the vascular system of both kidneys.

Thus, a multicystic kidney discovered prior to or during an operation is subject to removal. Postoperative prognosis and remote results are quite favourable.

SPONGE KIDNEY

This is a peculiar cystic affection of the renal pyramids. It is characterized by multiple small cysts in the pyramids. In the literature this disease has several names. Among them are multicystosis of the medullary substance, congenital cystic dilatation of the collecting tubules of the kidney and medullary sponge kidney. Genarduzzi was the first to use the term sponge kidney in 1939 and it is currently accepted by practically all authors.

Many research works, mainly by Italian authors, have lately been devoted to this cystic disease of the renal pyramids. They are all mentioned in Cachi's (1960) monograph on the sponge kidney. Thanks to this research, the disease began to be diagnosed much more often in recent times.

Abeshouse and Abeshouse (1960), for instance, collected 131 cases of sponge kidney from the world literature and added five of their own cases. Palubinskas (1961) published 29 cases, while Secrest and Kending (1961) described three cases where the diagnosis was made by means of X-ray examination. Murphy et al. (1961) described another seven cases of diagnosed sponge kidney.

In the Soviet literature Dzhavad-zade (1964) reported a case of a sponge kidney in an 8-year-old child (Fig. 15) and later we encountered it in another six adult patients and two children.

The aetiology, clinical aspects and treatment of sponge kidney were also discussed in the works of Strauss (1962), Gabriele (1964), Steyn and Logie (1964), and others.

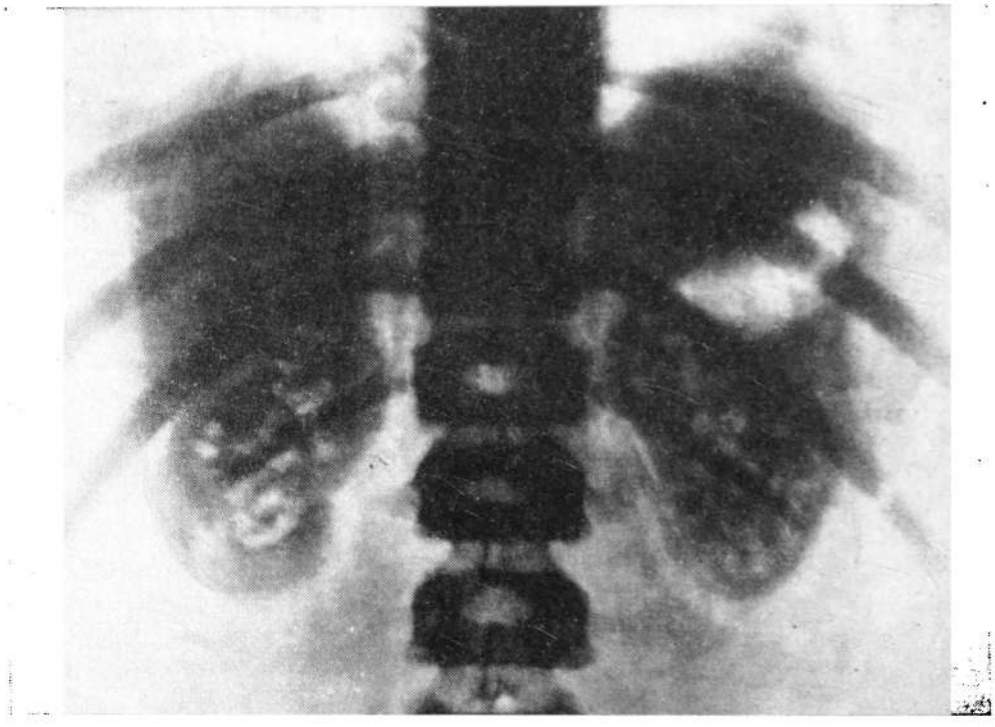


Fig. 15. Sponge kidneys complicated by nephrolithiasis (scout urogram)

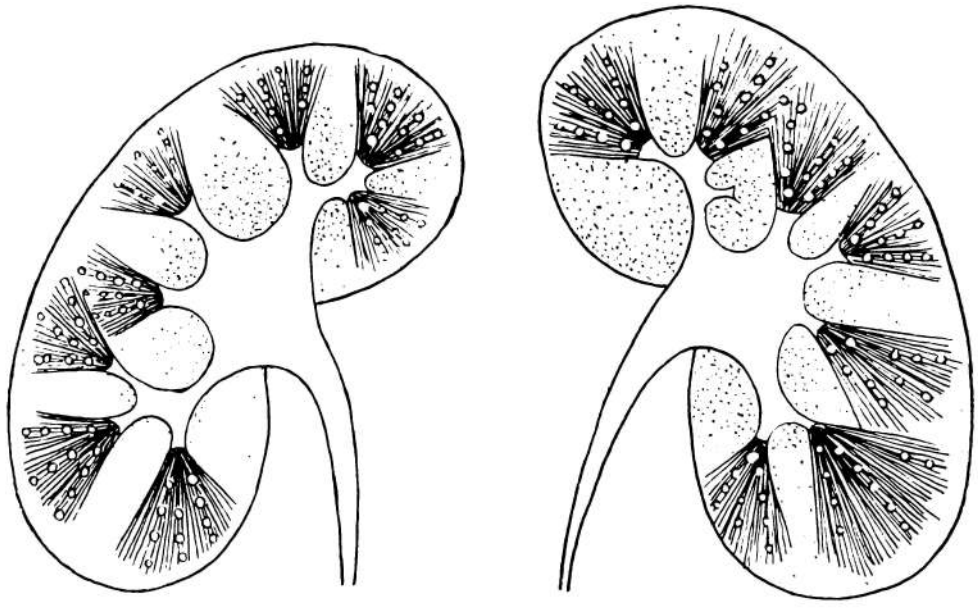


Fig. 16. Sponge kidneys (diagram)

In this form of cystic affection the kidneys are normal in colour and their surface is smooth. On section a vast number of tiny cysts or semispherical small cavities ranging from a pinhead to 3 mm in diameter are found, exclusively in the zone of the pyramids and without extending to the renal cortex. Some cysts protrude into the cavity of the calyces in front of the papillae. The cysts are irregular, and most often radially scattered over the whole surface of the pyramids, presenting the appearance of a sponge, from which the disease takes its name (Fig. 16). Small stones or sand, most often of a dark colour, are contained in the cavities of some of the cysts.

The cortical layer of the kidney is somewhat thinner than normal. Bertin's columns are slightly compressed between the pyramids. The mucosa of the pelvis and calyces is normal in appearance and usually unchanged.

In most patients the disease is bilateral. It is predominantly men that fall ill. The main symptoms of the disease are: haematuria, lumbar pain, renal colic and pyuria. Haematuria occurs in 85 per cent of cases and may be macro- or microscopic. The cause of renal colic may be stones that rupture the wall of the cyst and become lodged in the pelvic cavity. The course of the disease may be complicated by infection causing micturition disorders. The functional ability of the kidneys remains normal for a long time. With the onset of infection, renal function may be disturbed if the upper urinary tract becomes obstructed. The diagnosis of sponge kidney is based on X-ray examination. Lindvall (1959) describes in detail the X-ray picture of the sponge kidney on the basis of studying 35 X-rays of patients suffering from this disease.

Fine calcification of the renal medulla is sometimes determined in scout X-ray examination. A whole series of tiny cavities are often seen in the medulla below the calyces in excretory urography. They look like the pores of a sponge and that is why the disease was named sponge kidney. Histological examination reveals no basic changes in the tubules and glomeruli, but there is usually a considerable connective-tissue reaction around the cystically changed pyramids. The cysts are usually small and numerous; they are located radially and only in the medulla corresponding to the pyramids and branch off from the collecting tubules. Sponge kidney is an anomaly of the embryonic development of the kidney, somewhat related in aetiopathogenesis to polycystic kidney, although in its clinical course and pathoanatomical picture it is in many ways different from polycystic disease of the kidneys.

Typical of the sponge kidney are:

(1) the uniform cystic affection of the kidneys;

(2) the small size of the cysts with a radial arrangement in the renal medulla, due to which the surface and size of the kidneys remain unchanged;

(3) the size of the cysts increases gradually toward the centre, usually reaching 2-4 mm in diameter. These cavities contain no products of secretion and are fenced off by a cobweb-like net in the form of a sponge. There are often mucoprotein drops within the epithelium or in the lumen of the cysts. Macroscopically the pelvis is normal.

Patients with this form of cystic anomaly of the kidneys require no treatment in uncomplicated cases. Treatment is usually applied in cases of total haematuria to arrest bleeding. Surgery is indicated only when life is endangered or, in cases of acute pyelonephritis, when non-operative therapy proves ineffective. When there are migrating concrescences which often obstruct the upper urinary tract, their removal by surgery is indicated.

In 1962 Strauss described the clinical and pathoanatomical picture of peculiar cystic disease of the renal medulla. The author analysed 18 cases of this disease that had been published in the literature. This disease begins with the gradual development of anaemia or marked salt exhaustion. Azotaemia often occurs in the absence of proteinuria or changes in the urinary sediment. Arterial pressure is usually normal. Fibrous osteitis is typical of young patients. Hyperplasia of the parathyroid glands and of the adrenal cortex, together with changes in the bones and bone marrow, are common findings at autopsy. The kidneys are usually smaller than or of normal size. The subcortical substance is filled with numerous small cysts and the structure of the cortical substance is completely disturbed. The remaining glomeruli are either normal or hypertrophied. The photomicrographs given in the work explain the pathophysiology of the disease. In the author's opinion the disease is caused by a developmental defect of unknown aetiology.

In the literature this disease has been confused with sponge kidney, with which it shares neither a clinical nor a pathoanatomical resemblance. The treatment boils down to the correction of salt exhaustion, acidosis and anaemia.

POLYCYSTIC KIDNEY DISEASE

Polycystic kidney disease is justly regarded to be the most severe developmental anomaly of the urinary system. This is because the process is bilateral and congenital in character. There is a great variety of opinions on the terminology, aetiology, pathogenesis, clinical picture and treatment of this disease in

the literature. Polycystic kidney disease occupies a prominent place among renal cystic diseases in incidence and the special features of its course.

Many clinicians distinguish the following types of the disease according to the morphological peculiarities and specific features in its clinical course: (a) polycystic kidneys in the newborn; (b) polycystic kidneys in children; (c) polycystic kidneys in adults.

Besides this, some authors distinguish polycystic kidney disease of either congenital or acquired character. Sanchez-Lucas (1951) also differentiates two types of polycystic kidneys. In the first type the condition is compatible with life, the metanephrogenic tissue being sufficiently developed in this case to allow the kidneys to function satisfactorily. In the second type the disease is incompatible with life; it is observed in the stillborn or newborn infants who live very briefly. Lambert (1947) distinguishes three groups of cysts in polycystic kidneys on the grounds of their anatomical localization: (1) glomerular, (2) tubular and (3) excretory.

The author contends that glomerular cysts are always closed and are found in the polycystic kidneys of the newborn. The tubular and excretory types of cysts are connected with the tubules and usually drain into the renal pelvis.

On comparing the results obtained by Lambert in his study of immature foetuses with the data in the latest works of Parkkulainen and other researchers devoted to the study of multicystic dysplasia of the kidneys, the conclusion can be reached that only a bilateral multicystic kidney and not polycystic disease of the kidneys can be the cause of a stillborn foetus.

As a rule, polycystic kidney disease is a bilateral ailment with a specific clinical and anatomical picture, identical both for children and adults. A unilateral true polycystic kidney is a rare disease. It is of congenital character in many patients. Two forms of polycystic kidneys are distinguished according to the clinical and anatomical picture: in the first, which occurs more often, the kidneys are quite large, their weight may together total 4000-5000 grams. In one of our cases giant polycystic kidneys weighing 9800 grams were found at autopsy. In the second form the kidneys are not enlarged; on the contrary, they are even smaller, as if they were hypoplastic. Obviously this depends to a large degree on the length of time and the severity of the inflammatory changes that have occurred in the polycystic kidney.

Terminology. This disease has different names: polycystic kidney disease, polycystic degeneration of the kidneys, polycystic nephrotubulodysplasia, and so on.

The name 'polycystic degeneration of the kidneys' given to

the disease is incorrect because it fails to reflect the essence of the process and gives rise to confusion. Numerous cysts in the kidney form not because of the 'degeneration' of the renal parenchyma, but because of disturbed kidney embryogenesis. The correct name for this disease is polycystic kidney disease.

Incidence. In 1928 Sokolov reported that polycystic kidneys were found in 192 of 50 198 autopsies. According to Bell (1950), polycystic kidneys were registered in 313 of 110 240 autopsies (1 : 352). In analysing the data from 10 734 autopsies Dalgaard (1957) found 24 cases of polycystic kidneys. According to the records of the Pathology Department of the First Moscow City Clinical Hospital (Arkhangelskaya, 1959), only 32 cases of polycystic kidneys were found in 9632 autopsies performed over a period of 12 years.

Gimpelson (1949) found polycystic kidneys in 51 out of 4549 patients with diseases of the kidneys and upper urinary tract. Pytel (1940) had six patients with polycystic kidney disease among 307 patients with renal ailments. Fransvea and Papadia (1959) note that polycystic kidneys account for 1.38 per cent of renal anomalies. The greatest number of cases in the literature were reported by the Mayo Clinic, with 336 patients with polycystic kidney disease over a period of 20 years (Simon and Thompson, 1955).

In our experience, from 1946 to the present day, there have been 276 patients with polycystic kidneys.

Analysis of the literature published during the last three decades shows that in adults polycystic kidneys were encountered once per 350-400 autopsies and in one patient per 150-200 urological in-patients. This was far more often than in former decades, most probably because diagnostics has improved so much.

Aetiology and pathogenesis. Polycystic kidney is justly considered to be the severest developmental anomaly of the urinary system. The first theory in explanation of its origin was offered over a hundred years ago by Virchow. In the time that has elapsed views on the aetiology and pathogenesis of polycystic kidneys have changed over and over again. Among the theories of aetiopathogenesis the following merit attention: (1) the inflammation-retention theory; (2) the neoplastic theory; (3) the syphilitic theory; (4) the theory of disturbed embryonic development of the kidneys.

The inflammation-retention theory was advanced by Virchow in 1855. At first Virchow thought that cystic changes in the kidneys were due to deposits of urate salts in the tubules, which caused obstruction and the formation of cysts. He later claimed that fibrosis and obstruction were the result of an inflammatory process which most often occurs in the area of the papilla (papil-

litis). Petterson (1903) and others in experiments on rabbits established that ligation of their renal papillae leads only to distension of the tubules and this is attended by atrophy. The kidney itself shrank but in no way resembled a polycystic kidney.

There are practically no advocates of the inflammation-retention theory at present.

The progressive growth of polycystic kidneys, resembling the growth of a tumour, caused some researchers to assume that a neoplastic process forms the basis of the disease. According to this theory, the development of renal cysts is of the adenocyst type, similar to adenocysts of the ovary, mammary gland and other organs. Karewski (1921) used the term 'cystadenofibroma' for the polycystic kidney. Later clinical follow-ups and histological studies ruled out the neoplastic theory of the origin of the polycystic kidney disease.

The role of congenital syphilis in the origin of polycystic kidneys was emphasized by Telyatnikov (1936) and other authors who regarded polycystic kidney disease to be a consequence of congenital syphilis and even suggested specific treatment for such patients. In our opinion, the views of these authors on the aetiopathogenesis of polycystic kidney disease cannot stand up to criticism. None of our patients appeared to have any history of syphilis. This opinion is shared by many other authors. The so-called syphilitic theory of the origin of polycystic kidneys is rejected by most researchers.

The theory meriting the greatest attention is that of disturbed embryonic development of the kidneys. Many works are devoted to the elaboration of this theory, among which special interest should be given to the publications of Norris and Hermann, 1941; Lambert, 1947; Sanchez-Lucas, 1951, and others. Acquaintance with these publications permitted us to establish that the studies described in them do support the theory of disturbed embryonic development of the kidneys in the genesis of policystic kidney disease. Most researchers think that its aetiopathogenesis results from the improper development of the kidney in ontophylogenesis consisting in the untimely and abnormal fusion of its excretory and secretory apparatuses.

To clarify the pathogenesis of polycystic kidneys Norris and Hermann (1941) carried out scrupulous histological studies of such kidneys both in infants and in adults. They were the first to establish that kidneys which later became polycystic had developed normally for a considerable length of time in the intrauterine period and it was only after the metanephrogenic blastema had differentiated and its segments displaced that cysts in the segments of different nephrons began to develop. These changes are degenerative in character, resembling the stage of normal de-

generation of the mesonephros and normal rudimentary metanephric elements. Abnormally preserved elements of the metanephros in various stages are often encountered in polycystic kidneys. This explanation fits most congenital anomalies connected with polycystic kidneys and corresponds to the congenital character of the disease.

The theory advanced by Kampmeier (1923) was based on the results of studying polycystic kidneys of a human embryo. Kampmeier described three limited zones in kidney development: the 'rudimentary' zone, the 'temporary' zone, and the 'growing' zone. The first generations of the urine-excreting tubules comprise the 'rudimentary' zone. These tubules do not communicate with the first generations of collecting tubules and disappear without a trace. The 'temporary' zone consists of three subsequent generations of metanephric tubules, which communicate with the corresponding generations of the collecting tubules; then they close to form, by the third month of intrauterine life, closed metanephric vesicles. In the following weeks in normal embryogenesis these vesicles become atrophied and disappear. The following generations which form the permanent kidney, while forming along the periphery of the 'temporary' zone, create the 'growing' zone.

According to Kampmeier, in a polycystic kidney the vesicles of the 'temporary' zone grow instead of undergoing progressive atrophy. The developing 'temporary' zone compresses the normal collecting tubules and promotes the formation of cysts. Although this theory explains the origin of polycystic kidneys from a mechanical viewpoint, it nonetheless deserves attention. It helps to explain the formation of cysts in the medulla of the kidney; but in a polycystic kidney the cysts are widely scattered both in the medulla and the cortex. To this conclusion also arrived Andrieu (1958). On studying in graphic reconstruction the kidneys of two newborn babies and five adults with polycystic disease, Lambert (1947) reached some original conclusions. On sections of adult polycystic kidneys he found, as has been mentioned above, three types of cysts irregularly distributed in three zones:

(1) large glomerular cysts were located immediately under the renal capsule;

(2) smaller tubular cysts were located in the deep cortical layer; they were filled with a dense homogeneous coagulant like that which is found in the distal ends of tubules in patients suffering from proteinuria;

(3) small excretory cysts were located in the renal medulla. Being continuous with the collecting tubules they are found on serial sections to the point where the tubules drain into the calyx.

Without denying the role of disturbed kidney embryogenesis

in the development of polycystic disease, Lambert concludes that the genesis of this ailment remains unclear. Lambert's research into the function of the cystically changed nephron is very valuable.

Bialestock (1956) takes a new point of departure in his attempt to explain the aetiology of polycystic kidney. She contends that the disease is an example of epigenesis; in this condition embryonic tissues do not develop according to a predetermined pattern, but according to the pattern that is most suitable for their environment. It is possible that such pathological stimuli (hormones, chemical substances, etc.) will be found in the future which cause abnormal conditions in the environment and directly in the nephron itself or in the organism as a whole, with a subsequent effect on the nephron. According to Bialestock, whatever this pathological factor might be, the earlier its effect is felt the longer it continues, and as a result of this the kidney is even more distorted. In polycystic kidney the whole renal blastema that originated from the mesonephros and the mesonephric duct may be affected.

It is worth recalling that polycystic kidneys occur in some birds and fish. In this regard the studies conducted by Schlumberger (1950) are of some interest. He surveyed the basins where goldfish were bred and found that 5 per cent of them are born with polycystic kidneys. Four such goldfish were examined in great detail and the functions of their kidneys closely studied. The nitrogen content of their blood was twice that of the blood of fish with normal kidneys. The goldfish with polycystic kidneys discharged half the urine a normal fish excreted.

Summing up all that has been said about the aetiopathogenesis of polycystic kidneys, it can be stated that of all the proposed theories the most acceptable is the theory of disturbed embryonic development of the kidneys. Most morphologists and clinicians now adhere to this interpretation of the origin of renal polycystic disease. It is a view with which we agree.

As was mentioned above, normally the complete joining of the straight and convoluted tubules occurs during the embryogenesis of the kidneys, in other words, there is proper formation of the nephron.

In polycystic kidney the nephron is formed improperly because the straight and convoluted tubules of many nephrons are joined incompletely and incorrectly during embryogenesis. This abnormal fusion of the excretory and secretory systems makes the drainage of urine (filtrate) from the proximal parts of the nephron difficult. As a result of increased pressure, the tubules proximal to the obstruction dilate and small cysts then form. If the straight and convoluted tubules do not join at all, then large cysts are formed. This is why there are usually several large cysts in poly-

cystic kidneys along with numerous smaller cysts located on the surface of the kidneys. In the progressive postembryonic cystic changes an important role is also played by involvement of the polycystic kidneys in pyelonephritis. The chronic inflammatory process causes the proliferation of interstitial tissue and compression of the tubules, which in turn hampers still more the flow of urine in the cystically changed nephron. This ultimately leads to the death of the nephron and the formation of large cysts.

It is essential to note that there may be properly formed nephrons along with malformed ones. This explains the presence of areas of normal renal parenchyma between the cysts in the early stages of polycystic kidneys which are not affected by pyelonephritis. This is verified by data from histological examinations which show that, together with malformed glomeruli and nephrons, normal glomeruli and tubules are also found in the specimens.

Both the data in the literature of the last few decades and our own studies have confirmed the maxim that a true polycystic kidney disease is always bilateral. The cysts in one of the kidneys may, however, be so small that they cannot be identified even by the latest modern diagnostic techniques.

Lambert (1947) studied the glomerular and tubular function of cystically changed nephrons by injecting inulin into the abdominal cavity of patients a few hours before their death and then studying the contents of the cysts for the presence of inulin, urea, creatinine, glucose, etc. These studies revealed that the cystically changed nephrons of adults had to a considerable extent preserved their functional ability and participated in the formation of urine. The same conclusion was reached by Patton and Bricker (1954).

Some investigators examined the contents of the cysts and urinary bladder. Making a comparative study of the fluids they came to the conclusion that the content of the cysts is not a simple fluid transudate, but urine produced by filtration through the glomerulo-tubular system of the kidney.

These studies which reveal that the cystically changed nephron is a functional unit are of much practical value, particularly in surgical operations. It becomes therefore clear that in performing ignipuncture care should be taken, if possible, not to destroy cysts smaller than 1-1.5 cm.

Very often patients with polycystic kidneys also have a polycystic liver, which is explained by the close functional and morphological correlations of these organs in the process of embryogenesis. The frequently noted hereditary character of this disease also convincingly shows that polycystic kidney is a congenital renal anomaly. Study of the role of heredity in the genesis of this disease is a very complex and difficult task. In collect-

ing the medical history thoroughly one often finds that relatives and members of the patient's family have polycystic kidneys. This has prompted many authors to advance the theory of the role of heredity in polycystic kidney disease.

An important work was written by Morgani in 1949 (cited from Dzhavad-zade, 1964) who studied the medical histories of 169 blood-relatives of patients suffering from polycystic kidneys in six generations; 95 of them were clinically examined. The author established that the heredity is dominant, direct and monogenic in character with transmission in one case by direct descent through four generations. Higgins (1952) also points to the hereditary factor in the origin of polycystic kidney disease. He examined the family members of 94 patients and found that those of 34 patients also had polycystic kidneys. Higgins concludes that in the diagnosis of polycystic kidney disease the role of the hereditary factor is one of the most prominent.

In the Soviet literature Shapiro (1928) and Gotlib (1925) emphasized the role of the hereditary factor in the genesis of the disease.

It should be noted that over the years many works have been devoted to the role of the hereditary factor in the disease (Dalgaard, 1957; Würschung, 1957, and others). The most basic of these studies was the research carried out by Dalgaard (1957), who summed up his own numerous studies of polycystic kidney among the members of one family through several generations.

We studied 223 family members of 83 patients with polycystic kidneys by means of personal contact with relatives. We collected information about deceased relatives and took excerpts from medical histories, death certificates, etc. Father, mother, brothers, sisters and children were subject to investigation; of 48 members of the families 46 patients proved to have had polycystic kidneys.

In examining the members of families of patients our attention was drawn to the fact that 36 patients had inherited the anomaly along the maternal line. One family is particularly interesting in this respect: two sisters were operated on for polycystic kidneys; their mother died from the same disease when she was fifty, while the third sister is currently in our care, also with polycystic kidneys following an uncomplicated course.

The most difficult part of the problem of heredity lies in ascertaining the regularities and character of the inheritance of polycystic kidneys in different generations.

The problem cannot be solved by using the Dalgaard formula to count mutations in this anomaly.

We know that the process of evolution is based on changes in the reproduction of the parents' characteristics in their prog-

eny. They are called mutations. To some extent polycystic kidneys possess the features of mutations. The correct determination of the frequency of polycystic kidney mutations and the possible control over the qualitative side of mutations to change their initial morbid character are tasks to be solved in the future. At present, however, research into the questions of the role of the hereditary factor in the origin of this disease is limited to the statement of clinical and statistical data. It should be emphasized that the data obtained by such research are of immense practical significance in the diagnosis of polycystic kidney disease.

In summing up all that has been said regarding the role of the hereditary factor in the genesis of polycystic kidneys, it is essential to emphasize again the hereditary character of the disease. One must accept the views of those authors who attach much diagnostic importance to this and who recommend examining all the members of the family of a patient with this kidney anomaly. This is also of immense prophylactic importance because early diagnosis and timely treatment prevent many complications and the progress of the disease.

Pathology. There are only a few works devoted to the special study of the pathology and the histological structure of polycystic kidneys at different stages of the disease. In the Soviet literature we found no special research into the problem. Only Sokolov (1927) partially describes the histological picture of this malformation in a patient with a polycystic kidney and liver, while Arkhangelskaya (1959) for the most part compares clinical and anatomical diagnoses in cases of polycystic kidneys.

In his classic manual on the pathology of renal diseases Allen (1951) describes the histological picture of a polycystic kidney without dwelling on the role of pyelonephritis in the pathogenesis of renal insufficiency and without comparing the results of morphological examinations with clinical data. The pathohistological data that are provided are of a purely general character.

There can be no doubt that only the systematic study of a rich fund of clinical and pathological material can provide the key to many complex questions of the aetiopathogenesis and clinical treatment of this renal anomaly.

In comparing the clinical and morphological data one should clarify the pathogenesis of renal insufficiency, developing mainly as a consequence of pyelonephritis. This gives rise to another problem: how does the infection spread? The development of hypertension in most patients with polycystic kidneys and the frequently fatal outcome of cerebral haemorrhage sets us the task of clarifying the connection between arterial hypertension

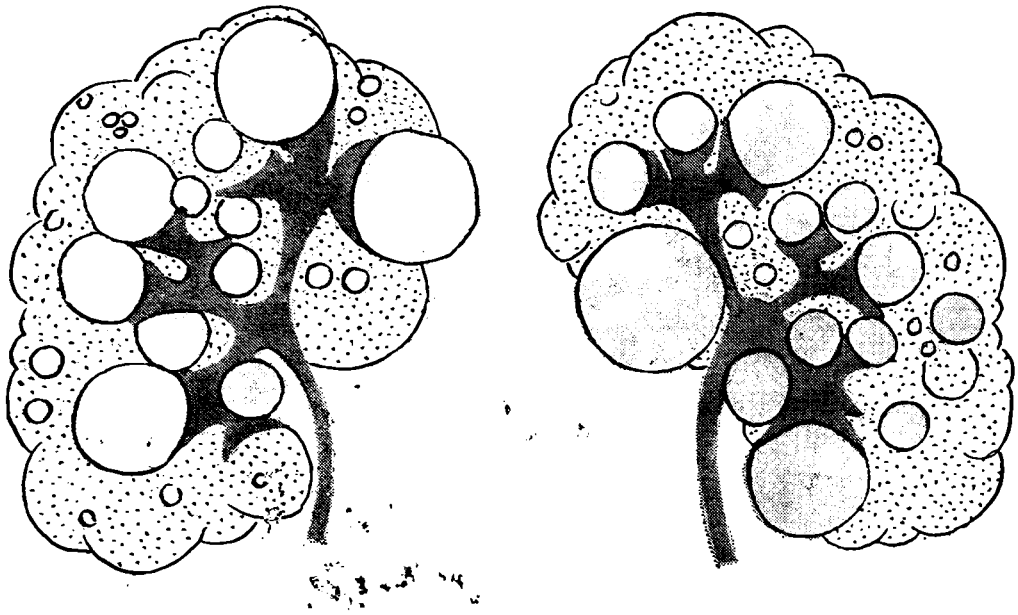


Fig. 17. Polycystic kidneys (diagram)

and morphological changes in the polycystic kidney and cerebral vessels.

We studied the findings of 29 autopsies and the biopsy material of 32 patients who had been operated on. The results of the morphological examinations were compared with the data of clinical follow-ups and treatment of 276 patients with polycystic kidney disease.

Morphology. Polycystic kidneys are extremely variable in shape and size (Fig. 17).

Figs. 18 and 19 illustrate this. The characteristic feature of all polycystic kidneys, however, is the presence on their surface and in their parenchyma of a large number of cysts of various sizes containing fluids of different colours. The size of the cysts in small polycystic kidneys rarely exceeds 1-3 cm and section reveals a multichamber structure with a stroma consisting of thin layers of connective tissue. The renal pelvis, compressed by cysts, looks like a long fissure.

A section of these kidneys shows evenly positioned cavities of different diameter resembling irregular bee hives. On the surface of large polycystic kidneys one can see quite large cysts (up to 6-8 cm in diameter) together with small cysts. The larger cysts usually localize at one of the poles, or more rarely in the depth of the parenchyma and at the hilus. In the latter case these cysts may cause haemodynamic disorders in the kidney; it is quite possible that they cause the development of nephrogenic hypertension.

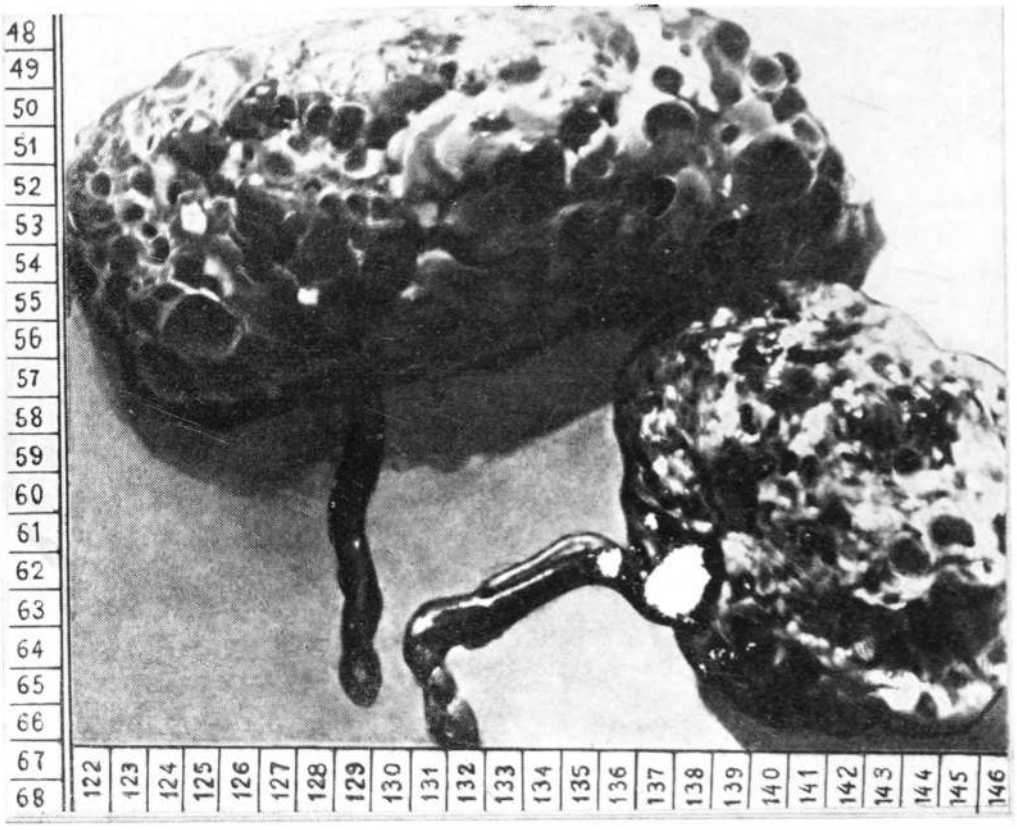


Fig. 18. Polycystic kidneys removed after death from the body of a 48-year-old woman. Death occurred from haemorrhage into the brain after the rupture of a congenital aneurysm of the middle cerebral artery

There is usually an intermediate layer of renal parenchyma between the cysts. The amount of cystically unchanged parenchyma depends on the presence and duration of the process. The colour of the parenchyma of such a kidney is either normal or with a greyish hue. The general appearance of the polycystic kidney is often compared to a bunch of grapes. The fluid in the cystic cavities is usually clear; when there is haemorrhage in the cyst the fluid becomes brown, whereas in inflammation a purulent fluid appears. The cysts that are not suppurated are filled with an aqueous fluid containing elements of urine (urea, uric acid, salts, cholesterol); in other cases the contents are a jelly- or pastelike mass, as a result of the presence of colloid substances: fat, epithelium, detritus, erythrocytes. Microscopic examination of the contents of non-suppurated cysts reveals single leucocytes, erythrocytes, epithelial cells, spherical hyaline corpuscles, and crystals of uric acid or calcium oxalate.

Microscopy of the cysts reveals that their inner surface is lined

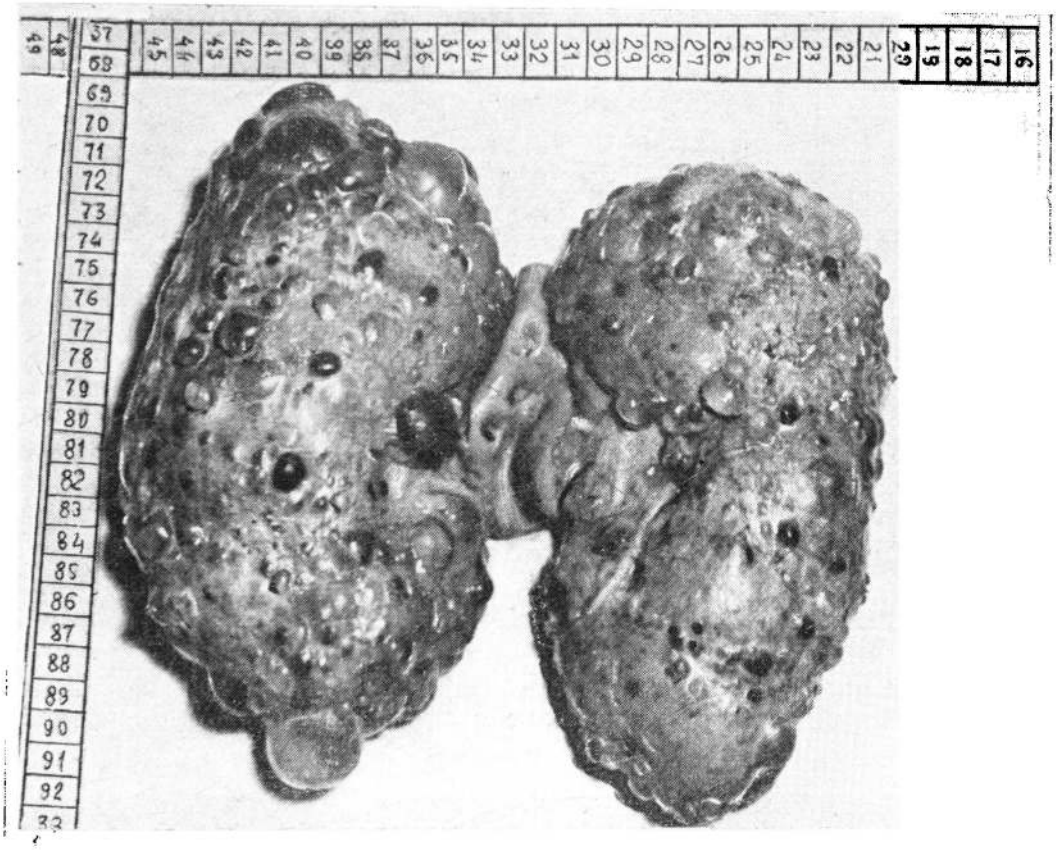


Fig. 19. Polycystic kidneys removed from a 62-year-old woman who had died from uraemia

with a cuboidal, sometimes flattened epithelium. In larger cysts a considerable length of the epithelium can be desquamated. Quite often the epithelial elements proliferate in the form of polyposis, which is obviously connected with the chronic inflammatory process in the wall of the cyst. Most of the cavities are usually filled with a homogeneous glassy mass which stains an intense violet with haematoxylin. With suppuration leucocytes accumulate in the protein content of the cysts. The walls of the cysts consist of a very thin layer of dense connective tissue which is the only septum between big cysts in some places. In some specimens we have discovered remnants of the renal parenchyma in these septa. The parenchyma around the smaller cysts is preserved to a greater extent. The urinary tubules in bioptic specimens are usually in various stages of distension. In some cases it is possible to trace on a series of specimens the various degrees of transition from the normal lumen of the tubules to dilated tubules and to note the moderate clustering infiltration of the stroma by lymphocytic cells.

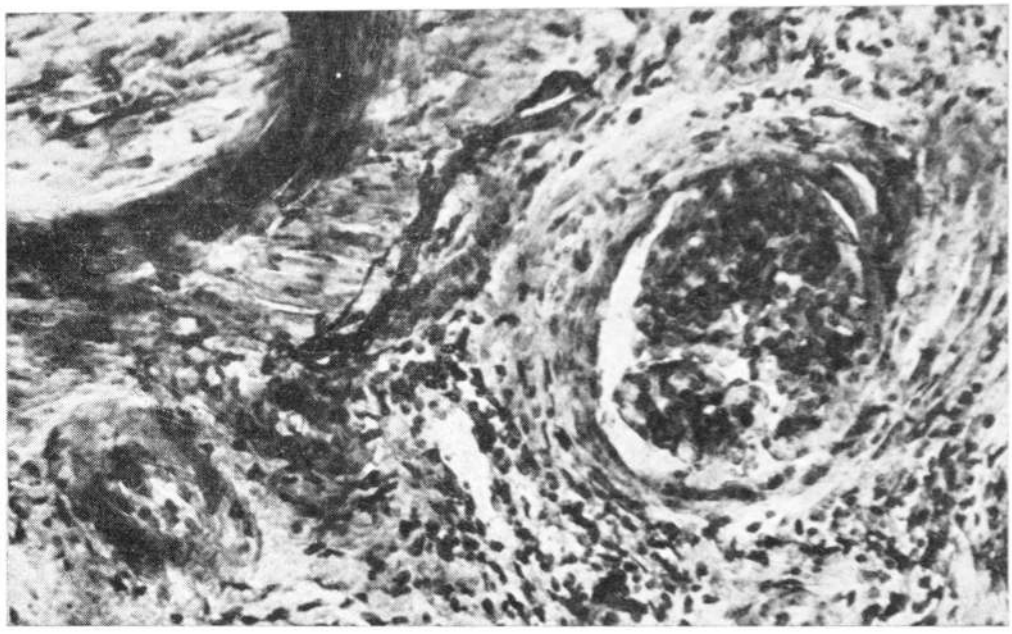


Fig. 20. A nerve bundle consisting mainly of non-medullated fibres, seen in the wall of the cyst among the smooth muscles. (Campos' method)

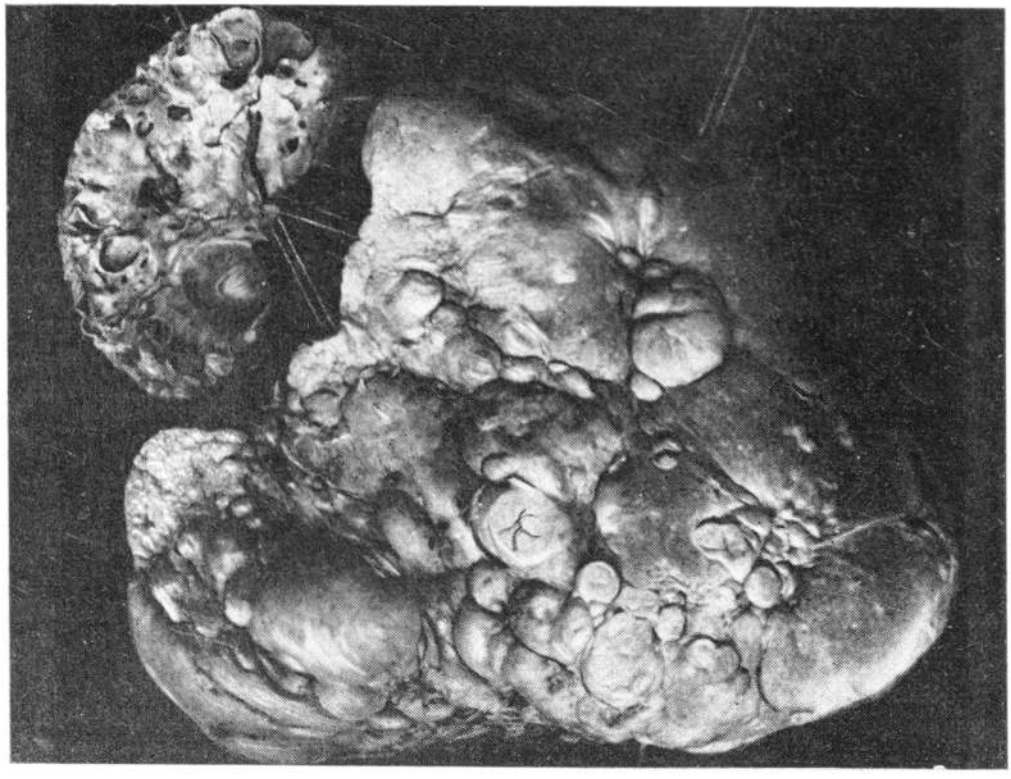


Fig. 21. Combination of polycystic kidney and polycystic liver. Death occurred from renal insufficiency, uraemia, and bilateral pyelonephritis

Nerve bundles with argentophil fibres were found around the large arteries in the renal tissue; this is the sign of the initial degenerative changes in them. The nerve bundles consisted mainly of non-medullated fibres with a small admixture of medullary fibres. Small nerve bundles of non-medullated fibres stretched mainly in the smooth muscles in the walls of small cysts (Fig. 20). Non-medullated nerve fibres were also seen in the tubules. The fine medullated fibres in the walls of the cysts were marked by degenerative changes manifested in increased argentophil properties and even the disintegration of some fibres. In most specimens we found the nerve fibres in the zones of the most severe inflammatory changes. We discovered these changes in specimens of bioptic material taken from patients operated on in the compensated stage of the disease.

In studying the nerve structures our attention was drawn to the fact that in polycystic kidneys quite a small number of nerve elements are found, although the degree of impregnation of these visible nerve bundles and fibres is considerable. This suggests that in polycystic kidneys with 86 per cent pyelonephritis affection nerve elements disintegrate and together with other tissues die quite rapidly. It is our opinion that the death of the nerve elements in a polycystic kidney is due first and foremost to anoxia consequent upon ischaemia of the renal tissue.

The **combination of polycystic kidneys with other anomalies.** As we have already pointed out, polycystic disease of the kidneys can be combined with anomalies in other organs. The combination of anomalies is often discovered in autopsies. In most children who had polycystic kidneys and died, the anomalies of other organs were too severe to be compatible with life. In adults the combination of polycystic kidneys with other anomalies compatible with life can be encountered at any age. Sometimes the anomalies of other organs may be the cause of various morbid processes, even ending in death. Polycystic kidneys are often combined with polycystic liver, pancreas, spleen, lungs, ovaries, and also with aneurysm of the cerebral arteries, and so on.

Among the concomitant anomalies polycystic liver is most often encountered. This has already been pointed out by other authors. Thus, for instance, Sokolov (1928) found that among 192 patients with polycystic kidneys 39 also had a polycystic liver; out of 173 autopsies Dalgaard (1957) found 76 cases with both polycystic kidneys and a polycystic liver; among 46 autopsies Rall and Odel (1949) registered such a combination in 15 cases. The combination of polycystic kidneys with polycystic liver can be explained by the ontophylogenetic link between these two organs, the close functional ties of which also exist in the postnatal period. Among 29 autopsies we found 11 cases with

polycystic kidneys and concomitant polycystic liver (Fig. 21).

We observed the combination of polycystic kidneys with polycystic pancreas in two cases. Dalgaard (1957) never encountered this combination of anomalies, whereas Sokolov (1928) had two cases with polycystic pancreas among 192 autopsies, and Rall and Odel (1949) recorded such a combination in four cases out of 46 autopsies.

We encountered polycystic spleen and polycystic ovaries only. once. A similar combination of polycystic kidneys, spleen and ovaries was encountered by Rall and Odel, while a cyst of the ovary alone was observed by Sokolov and Aleksapolsky (1923).

Among 29 autopsies we found one case of polycystic kidneys and aneurysm of the middle cerebral artery, where death was caused by cerebral haemorrhage. That we observed this just once is evidently explained by the fact that formerly the brain of patients who had died of polycystic kidney disease had been examined without taking into account the possibility of there having been an aneurysm of the cerebral arteries, because the data in the literature record this combination of anomalies more often. Suter (1949), for instance, recorded five patients with aneurysms of the cerebral arteries and concomitant polycystic kidneys, while Poutasse and Cardner (1954) three such patients, all young people (under 30) who had been admitted to the clinic for subarachnoidal haemorrhages.

On summing up the data of 11 245 autopsies, Brown (1951) established that polycystic kidneys were found in 36 cases and cerebral aneurysms in 148; six of the patients with polycystic kidneys had cerebral aneurysms and all six died from subarachnoidal haemorrhages. Sahs (1950) described ten cases of intracranial aneurysms discovered at autopsy, in four of which there were also polycystic kidneys.

One of our patients with polycystic kidney disease who died from uraemia had an aneurysm of the splenic artery revealed on autopsy. Macro- and microscopically this aneurysm was arteriosclerotic with areas of calcification. This is a rare phenomenon and we could only find one other such case in the literature. Harrow and Sloane (1960) described a case where a patient had the following diagnosis: polycystic kidneys, aneurysm of the left renal artery, aneurysm of the splenic artery, stone in the right kidney, and hypertension.

Among our cases we had one in which we discovered at autopsy a polycystic horseshoe kidney, both halves of which were uniformly affected by cysts. Such a combination of two anomalies in the kidneys is very rare. Sokolova (1954) described a case of nephrectomy of half a polycystic horseshoe kidney. A similar operation was performed by Vorobtsov (1956) for total haema-

turia. Carlson (1946) and Bolgarsky (1961) each described one case of an extremely rare combination of anomalies: polycystic disease of a congenital solitary kidney. An interesting case was described by Allansmith (1958). A baby that died soon after birth had a solitary polycystic right kidney with ectopia of its ureter which drained into the seminal vesicle.

The combination of polycystic kidney disease with multiple cysts in the lungs has been described by Bocian and Liang (1955).

A rare combination of anomalies of the urinary system was found in three of our patients: duplication of the upper urinary tract on one side and polycystic kidneys. A similar case was reported by Aivazyan (1961).

Summing up the data of our pathological studies and analysing the available literature it is essential to state that polycystic kidneys are a congenital disease. One of the main causes of disturbed normal balance in the organism of patients with polycystic kidney disease, and also of a fatal outcome, is renal insufficiency. At the same time one of the major factors promoting the origin and progress of renal insufficiency in polycystic kidney disease is pyelonephritis, which remains latent over a long period and only after a number of years, by producing periodical aggravations, manifests itself clinically. The peculiarities of disturbed circulation in the kidneys and impaired flow of urine, as well as the specific histological structure of polycystic kidneys, are conducive to this.

Thus, it is the infection of the cysts and the development of pyelonephritis, rather than the presence of a large number of cysts in the renal parenchyma, which are a threat to the patient's life. This is the key point to be considered both in the diagnosis and treatment of polycystic kidney disease.

Infection enters and spreads in the renal parenchyma by two routes, urinogenous and haematogenous. The haematogenous route evidently prevails in polycystic kidney disease. The causative agents are usually *Escherichia coli* and staphylococcus.

Pyelonephritis is encountered in 86 per cent of cases of polycystic kidneys and is usually bilateral. The nephrogenous character of hypertension in patients with polycystic kidneys is verified by histological findings in the vessels of the affected kidneys. The pathogenesis of the frequently observed arterial hypertension in these patients is evidently similar to the pathogenesis of hypertension in patients suffering from chronic pyelonephritis. Therefore, in choosing the method of treatment for hypertension in patients with polycystic kidneys, it is necessary to prescribe, besides symptomatic therapy, measures for the management of pyelonephritis.

In addition to circulatory disorders a frequent cause of hae-

Fig. 22. Horizontal Kristeller section through the middle of the renal pelvis. Leucocyte aggregations, found mainly in the region of the papilla, are stained black. Purulent melting of the papilla with suppuration spreading in the interstitial tissue and under the capsule. Secondary suppuration of cysts. Intense leucocytic infiltration of mucosa and submucosa of the renal pelvis. Goldman's stain. The specimen was prepared after death from the polycystic kidney of a 46-year-old man

maturia in polycystic kidneys is necrosis of the papillae and the fornical zones of the calyces. We found such changes in examination of Kristeller sections (Fig. 22).

Symptomatology. Polycystic kidney disease has a variety of

manifestations related to the patient's general condition and local symptoms.

The general symptoms include rapid fatiguability, cardiovascular abnormalities (increased arterial pressure, pain in the region of the heart, retrosternal pain), pain in the epigastrium, and thirst, while the local manifestations are pain in the small of the back, disorders of micturition, and a palpable tumour.

Rapid fatiguability is often due to pyelonephritis. In such cases the patients usually complain that they feel jaded and indisposed by the end of work. Such manifestations are observed for quite a lengthy period of time. Rapid fatiguability and malaise are particularly marked in patients in whom polycystic kidneys are complicated by pyelonephritis and renal insufficiency.

Dull pain in the small of the back is the most common, though not pathognomonic, symptom of polycystic kidney disease. It is due to: (1) disturbed flow of urine; (2) circulatory disorders in the kidney; (3) inflammatory processes in the kidney.

The mobility of the kidney, the progressive increase in the contents of the cysts in haemorrhage into them, the presence of concrements in the urinary tract, etc. promote to a large extent the development of lumbar pain. More or less intense pain is felt by the patient to the end of his life.

Cardiovascular disorders are manifested by pain in the region of the heart, retrosternal pain, headache, and dizziness.

Arterial hypertension is also encountered in patients with polycystic kidneys. The arterial pressure ranges from 150/80 to 220/110 mm Hg. Pyelonephritis plays an important role in the pathogenesis of arterial hypertension. We discuss this problem in detail below.

It is important to note that in 20 per cent of cases cardiovascular disorders may be the only manifestation of polycystic kidneys, and it is because of them that the sick person seeks medical advice. Due to insufficiently careful examination of the kidneys a diagnosis of hypertensive disease is often made in this group of patients.

Oedema of the lower extremities is a rare cardiovascular symptom of the disease. Only 18 of our patients developed oedema at the severe or terminal renal insufficiency stage.

Thirst and polyuria accompany polycystic kidney disease quite often. These signs, thirst in particular, have characteristic distinctions. Thirst bothers patients in the summer particularly, when the weather is hot; daily fluid intake reaches four to five litres. This is quite understandable since the loss of water by the body through disturbed renal function must be compensated and the more so since copious sweating occurs in hot weather. The daily amount of urine excreted by patients ranges from 1800

to 3000 ml. It should be noted that thirst and polyuria are characteristic of polycystic kidney disease because the concentration of urine is disturbed early and frequently.

We encountered different degrees of renal insufficiency in 43 per cent of patients with polycystic kidneys.

Patients with marked and severe renal insufficiency, to say nothing of those at the terminal stage, have various gastro-intestinal disorders. Dyspeptic symptoms combined with pain are often attributed to a gastro-intestinal disease whereas actually their underlying factor is hyperazotaemia. With correction of hyperazotaemia the dyspeptic symptoms usually disappear to recur after different periods of time.

Morbid conditions of the respiratory passages, such as atrophic rhinopharyngitis, haemorrhagic laryngitis, etc., may be observed in renal insufficiency. They may be erroneously interpreted by the physician, which results in incorrect diagnosis and improper treatment. For instance, Karmanova and Levashov (1956) described a case in which haemorrhagic laryngitis called for tracheotomy; but the patient died from uraemia, which was a complication of polycystic kidney disease.

Incorrect diagnosis in 20 per cent of patients is also explained by the fact that the whole complex of symptoms of renal insufficiency usually develops only in the last stages. In the early stages the condition is usually latent and has unclear signs. It is our experience that renal insufficiency is often encountered in patients with polycystic kidneys, i.e. in 43 per cent of cases, and is frequently the last stage of the disease. A number of clinicians agree with this. For instance, Braasch and Schacht (1933) found renal insufficiency in 26 per cent of patients with polycystic kidneys and Dalgaard (1957) in 35 per cent of such patients.

Renal insufficiency in people with polycystic kidneys is manifested by general malaise, progressive weight loss, and anaemia. These symptoms are in direct proportion to the duration of the insufficiency. It was pointed out above that pyelonephritis is among the leading factors in the pathogenesis of renal insufficiency.

The *presence of a palpable tumour in the abdomen* may be the patient's only complaint. Because of their large size and mobility, polycystic kidneys can be palpated by the patient himself.

Macrohaematuria may be slight or very intense, transitory or protracted. Slight short-term macrohaematuria does not usually anaemize the patient, whereas when it is long-term and intensive it may cause severe anaemia and threaten the sick person's life. Such macrohaematuria may call for emergency surgery (nephrectomy or ignipuncture). Our three patients with total macrohaematuria were operated on because of its protracted and intense

character. Haematuria may be caused by impaired venous drainage from the kidney due to mechanical compression of the vessels in the renal hilus by a cyst. It would be right to assume that in such cases the bleeding occurs from the congested and varicose fornical veins of the kidney.

Macrohaematuria is a common complication of polycystic kidneys. Gimpelson (1949) encountered haematuria in 32 per cent, Rall and Odel (1949) in 23 per cent, Higgins (1952) in 41 per cent and Dalgaard (1957) in 45 per cent of patients suffering from polycystic kidney disease.

In most cases macrohaematuria is arrested by non-operative treatment (transfusion of blood, infusion of calcium chloride, Vikasol, vitamin C, bed-rest, etc.). Life-threatening bleeding from a polycystic kidney, however, is managed by surgery. Goldstein (1951) advocates ignipuncture in such cases. Galperstein (1929), Higgins (1952) and others performed nephrectomy. Vorobtsov (1956) undertook ignipuncture in a case with profuse haemorrhage, but this did not arrest the bleeding from a polycystic horseshoe kidney and heminephrectomy was then indicated and carried out.

The choice of operative method in profuse bleeding from a polycystic kidney depends, of course, on the cause of the complication as well as on the patient's general condition. Haematuria in our three patients was arrested by organ-preserving operations: two underwent ignipuncture and the third resection of the kidney. They all recovered.

Pain in the epigastrium may be caused by displacement of the abdominal organs by the large cystic kidneys and by considerably increased intra-abdominal pressure. It might also be due to gastritis or hepatopathy. An erroneous diagnosis is often made when patients complain of epigastric pain. Among our patients, for instance, two were treated for gastritis for several years and another two for hepatitis. It should be emphasized that these gastro-intestinal disorders are usually noted in patients with latent pyelonephritis, at the stage of chronic renal insufficiency.

Dysuria is not a characteristic symptom of polycystic kidneys: it may be due to secondary cystitis. Pollakiuria, caused by polyuria, which is often observed in patients with polycystic kidney disease, is a more frequent sign.

Clinical picture. Differentiation of the clinical course of polycystic kidney disease according to stages is of great practical importance not only in making the diagnosis and treating the patients, but also in prognosis.

Talman (cited from Dzhavad-zade, 1964) in 1929 suggested distinguishing three stages: the asymptomatic stage, the stage

with symptoms of renal insufficiency, and the stage with signs of compression of the adjacent organs and displacement of the kidneys.

Braasch (1917) also distinguished three stages: the latent stage, the stage characterized by enlargement of the kidneys and haematuria, and the uraemic stage.

Bell (1950) suggested distinguishing the early stage, when the diagnosis is made on the basis of palpation of the kidneys or characteristic changes in urograms; a middle, or surgical, stage when the kidneys are palpated, pain is felt and the body temperature elevated, and haematuria and suppuration of the cysts occur; and a terminal stage characterized by renal insufficiency.

Dalgaard (1957) also recognizes three stages: a stage when there are no symptoms characteristic of polycystic kidney disease; a stage when there are some symptoms of the disease; and the terminal stage marked by uraemia. Other classifications of the clinical course of polycystic kidney disease have been suggested but they have not found any adherents.

The main shortcoming of all the classifications suggested is that they are compiled with consideration only of functional changes in the kidneys. They do not reflect the role and condition of organs which perform the barrier function of the body and have no comparison of the clinical data with morphological findings, which is very important in making a prognosis of the disease. As a result, taking into consideration the stages suggested, the practicing physician will find it difficult to orient himself in determining the stage of the disease and choosing the methods of diagnosis and treatment.

From a comparison of the data of the clinical course of the disease with the findings of functional study of the kidneys, liver, stomach, salivary glands, cardiovascular system, etc., and from the results of histological examination of polycystic kidneys and bioptic material obtained during operation, Dzhavad-zade (1964) suggested the following three stages in the clinical course of polycystic renal disease: (1) compensation, (2) subcompensation, and (3) decompensation. We use this classification in our practice.

1. **Compensation stage.** The disease is usually characterized by dull pain in the region of the kidneys and general indisposition. In 20 per cent of our patients it was manifested by referred symptoms of ailments of the cardiovascular, nervous and alimentary systems. Elevation of body temperature was mostly encountered in females, during pregnancy in particular. In 50 per cent of patients arterial pressure was high, ranging from 170/80 to 220/90 mm Hg; the increase was usually transitory. There was no anaemia. Functional examination of the kidneys showed mild

diminution of their activity and irregularity in the disorder of renal function. Clinical symptoms of renal insufficiency were not observed. The functional condition of the liver, stomach, and intestine was quite satisfactory. In 20 per cent of patients it is difficult to make a diagnosis of the disease at this stage, but their condition allows the use of complex methods of X-ray diagnosis (angiography, presacral diagnostic pneumoretroperitoneum, etc.). According to our observations, if during this period there are no complications such as suppuration of cysts, frequent exacerbation of chronic pyelonephritis, total haematuria, etc., survival from the time that the first symptoms appear is 20 years on the average. At this stage patients are able to continue work. Surgical treatment is most effective at precisely this stage.

2. **Subcompensation stage.** Patients admitted to the clinic at this stage of the disease complain of pain in the region of the kidneys, thirst, a sensation of dryness of the mouth, occasional nausea, easy fatiguability, and headache. The course of the disease is marked by an undulant character, periods of exacerbation alternating with periods during which the patients' condition is satisfactory. The blood urea content ranged between 60 and 120 mg/100 ml. Anaemia of variyng degrees was found in all our patients. In 20 per cent of patients arterial pressure increased to 180/90 to 220/110 mm Hg. Various disorders of the cardiovascular system, gastro-intestinal tract, and liver were observed.

The working capacity of patients was drastically reduced.

3. **Decompensation stage.** Continuous nausea, at times with vomiting, general weakness, headache, and vertigo are the main complaints of patients at this stage. Dryness of the mouth and thirst troubled all our patients. There were frequent exacerbations of chronic renal insufficiency and pyelonephritis, for which reason the patients were repeatedly admitted to the clinic. Depending on the course of the disease the blood urea content ranged between 120 and 500 mg/100 ml. All patients suffered from anaemia and azotaemia. Functional examination of the kidneys revealed sharp disorders of both filtration and concentration capacity. Arterial pressure was increased in 18 per cent of patients from 190/90 to 220/100 mm Hg. All patients at this stage of the disease were incapable of working.

As to the course of chronic renal insufficiency, it should be noted that in polycystic kidney disease it usually progresses slowly with a gradual increase in nitrous residue in the blood although it is true that exacerbation of pyelonephritis sometimes interrupts the 'calm' course of the insufficiency. Another distinctive feature of chronic renal insufficiency in polycystic disease is that it remains asymptomatic for a lengthy period

of time (latent course). Quite often the whole drama of the disease breaks out at the terminal stage of insufficiency, which is marked by high hyperazotaemia and various complications of uraemia. A latent course of chronic uraemia in polycystic kidney disease is in turn due to the slow loss of renal functions, as a result of which the organism, evidently, can adapt itself better to the various disorders of homeostasis.

Diagnosis and differential diagnosis. The diagnosis of polycystic kidney disease is based on information gained from the medical history and on the results of general clinical, urological, and X-ray examinations of the patient. The diagnosis is clear when the disease is at the decompensation stage (azotaemia, anaemia, hypertension, etc.) and when palpation shows that both kidneys are enlarged and have a tuberous surface.

In most cases the diagnosis is unfortunately made late at the stage of marked or severe renal insufficiency. It is of course difficult to recognize polycystic kidneys when only one kidney is palpated or when both cannot be palpated and the clinical manifestations of the disease are similar to those of other illnesses (chronic nephritis and pyelonephritis, hydronephrosis, tumour of the kidney, etc.). It is very important to identify the disease in its initial stages since the condition is still being sufficiently compensated owing to which treatment aimed at preventing any possible severe complications is effective. If the disease is borne in mind, modern improved methods of laboratory, X-ray, and radioisotope examination will help the physician to make a proper and timely diagnosis in almost all cases.

Many clinicians attach much importance to the family history and the hereditary factor in the recognition of the disease. A properly collected medical history is essential in making a diagnosis in general, while in patients with polycystic kidneys it is extremely valuable. We encountered cases in which a polycystic kidney was found accidentally during operation, and only after surgery was additional information on the family history and the hereditary factor collected, which yielded data verifying the existence of the disease. If this had been done before surgery, polycystic kidneys would not have been a chance operative finding.

Palpation in the region of the kidneys helps to recognize the disease. Sometimes both kidneys grow to such a large size that protrusions are seen on the anterior abdominal wall. In bilateral enlargement the kidneys often differ in size.

During palpation it is very important to determine enlargement of both kidneys and any changes on their surface. Separate cysts are often palpated on the surface of both kidneys, which confirms the diagnosis beyond all question.

Changes in the blood in polycystic kidney disease depend mainly on the stage of the morbid process, the presence of an infectious focus in the kidneys, and the degree of azotaemia.

Laboratory analysis of the urine is not only of diagnostic importance, but is sometimes valuable in differential diagnosis. That is why with patients with polycystic kidneys urinalysis is repeated two or three times. Zimnitsky's test usually demonstrates hyposthenuria and hypoisosthenuria (1004-1010). This uniform level of the specific gravity of the urine was characteristic of many patients suffering from polycystic kidney disease. In most patients the urine has a low specific gravity, which is a consequence of damage inflicted to the tubules of the kidneys.

Proteinuria is usually slight and does not exceed 1 pro mille. Many authors recommend that physicians bear this in mind in differentiating chronic nephritis from polycystic kidney disease.

Besides routine urinalysis these days the Kakovsky-Addis and Nechiporenko's tests are also performed in cases of polycystic kidneys and pyelonephritis.

Pyuria occurs in 70 per cent of patients. Its high incidence indicates that polycystic kidneys are particularly sensitive to infection.

Hyaline and, especially, granular casts are rarely found in the urinary sediment. This circumstance is particularly important in differentiating polycystic kidney disease from chronic nephritis, rather than in making the diagnosis of the disease.

RENAL FUNCTION

In our practice we used the classification of Dzhavad-zade and Malkov (1975), which includes five stages of functional renal disorders: subclinical, initial, marked, severe, and terminal. Since each stage, with the exception of the subclinical stage, comprises periods of the disease it is possible to appraise not only the extent of renal function loss, but also the severity of the clinical course of the disease and the condition of the body barrier organs. A subclinical stage of tubular-interstitial type of renal insufficiency is characteristic of polycystic kidney disease. Among 43 per cent of patients suffering from renal insufficiency, half were at the subclinical stage with disturbed concentration of urine and reduced percentage of water reabsorption.

In the initial stage, polycystic kidney disease follows a favourable course; symptoms of functional renal insufficiency occur in the later stages and are often linked with inflammatory processes in the kidneys and upper urinary tract. Determination of the functional state of the kidneys and urinary tract in individuals with polycystic kidneys is important both for the diag-

nosis and prognosis of the disease. There are several methods of functional diagnosis of the kidneys. Some have already lost their initial importance. Others, though still important to some extent (alkali and urea loads, cryoscopy, etc.), are not strictly specific in renal diseases and are burdensome for the patients, particularly for those suffering from polycystic kidney disease.

We studied the functional state of polycystic kidneys in our patients by means of quantitative determination of urea in blood serum; Zimnitsky's test; clearance tests with sodium thiosulphate and cardiotrast; chromocystoscopy; excretory urography; radioisotope study; and renal angiography (in some patients).

Though these general clinical studies are important in the recognition of polycystic kidney disease, the decisive role in its diagnosis, however, still belongs to X-ray examination (retrograde pyelography, excretory urography, pneumoretroperitoneum, nephrotomography, etc.). X-ray examination of these patients, just as patients with various anomalies of the urinary tract, is of great practical value. The sequence of different types of X-ray examination of patients with polycystic disease and the results gained, however, differ greatly from those in other forms of kidney anomalies. There is hardly any other anomaly with an X-ray picture as diverse as that of polycystic kidneys.

The X-ray picture of polycystic kidneys reflects the pathoanatomical processes underlying this peculiar developmental anomaly. That is why the X-ray picture of the kidneys and upper urinary tract differs with the form of the polycystic kidney (giant or dwarfish), the extent of inflammatory and sclerotic changes in the kidney, and the size and localization of the cysts. Giant kidneys are encountered most frequently, small, or dwarfish, polycystic kidneys being less frequent.

With the gradual development of inflammatory and sclerotic processes in the kidneys, first small and then large cysts slowly grow. This alters the contours of the kidneys, which change not only in length but also in diameter. Their surface is irregular, macro- or micro-tuberous. The outlines of the calyces, pelvis and upper segment of the ureter also change. The progressive growth of the cysts and the intensification of sclerotic changes in the renal parenchyma and its vessels distort the angioarchitectonics of the polycystic kidney, which is manifested in the formation of avascular areas in the parenchyma (Fig. 23).

Scout radiography very rarely shows up shadows of an enlarged kidney. Tomography, diagnostic pneumoretroperitoneum or scanning of the kidneys yield much better results as regards the demonstration of the size and outlines of the kidneys.

Tomography (body section radiography) without the use of contrast media takes a more modest place in the examination

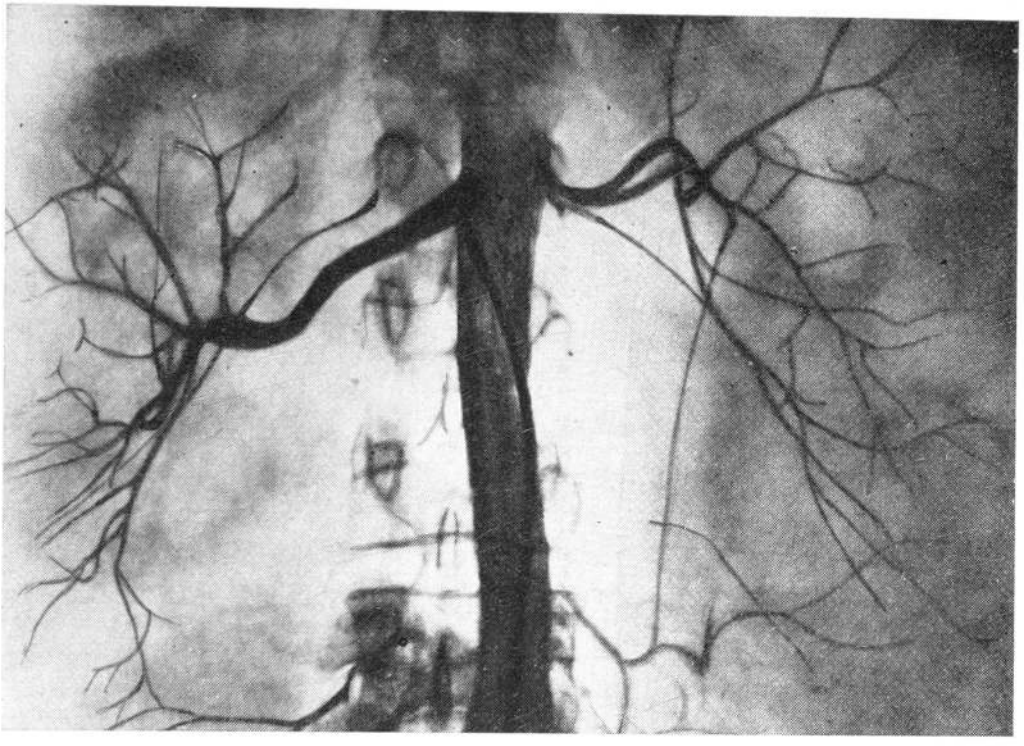


Fig. 23. Angiogram of polycystic kidney (12 months after an operation for omentorevascularization of the right kidney)

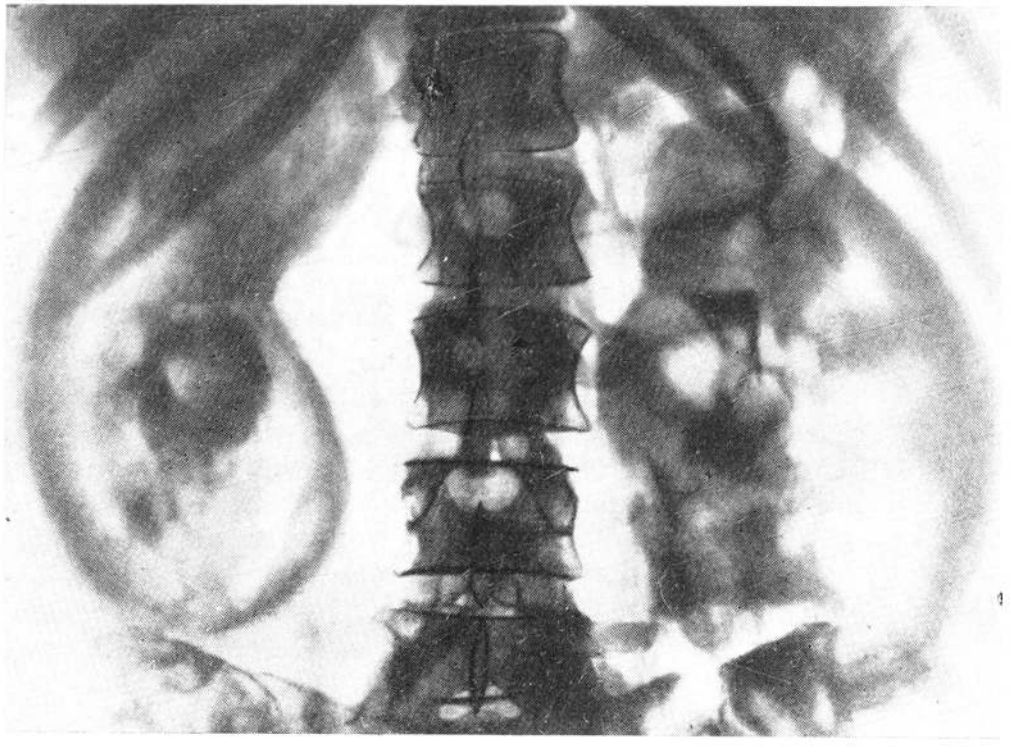


Fig. 24. Polycystic kidney disease. Pneumoretroperitoneum with excretory urography

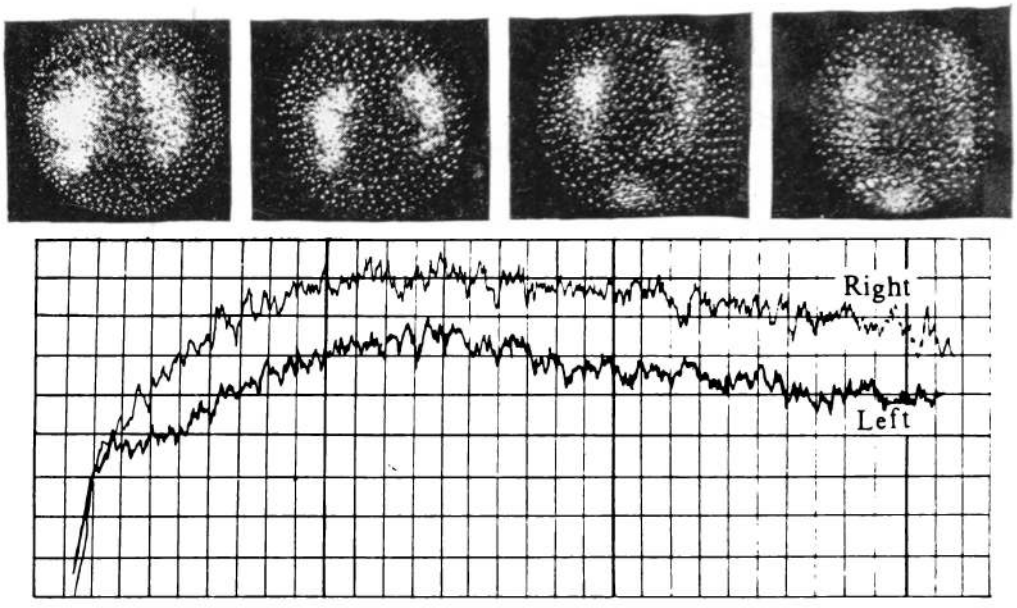


Fig. 25. Polycystic kidney disease. Dynamic scintigraphy and renography

of polycystic kidneys than in the detection of 'invisible' stones, since it is rarely able to show clear shadows of the kidneys. The results of presacral pneumoretroperitoneum are much better. A clearer picture is produced when diagnostic pneumoretroperitoneum is combined with excretory urography. In this case the radiographs show the enlarged outlines of the kidneys with an irregular surface and the presence of the contrast medium in the distorted pelvis and calyces (Fig. 24).

Lately, pneumoretroperitoneum is rarely used in the diagnosis of renal polycystosis, preference being given to scanning of the kidneys (Fig. 25) and aortography. When aortography cannot be carried out because of technical or other reasons and diagnosis of the disease is very difficult, pneumoretroperitoneum combined with excretory urography is quite expedient.

Retrograde pyelography is often applied to patients with polycystic kidneys. Asepsis and antisepsis should be properly ensured in such cases. The catheter should be advanced into the ureter for a distance of no more than 20 cm and at most 6 ml of the contrast medium should be introduced into the pelvis. From the findings of retrograde pyelography and general clinical examination, polycystic kidneys can be recognized in most patients. The diagnosis is verified absolutely if pyelograms show characteristic changes on both sides. It should be borne in mind that in the initial stages of the disease, in cases with 'dwarf' polycystic kidneys containing small cysts, it is very difficult to make the diagnosis based on the results of pyelography alone.

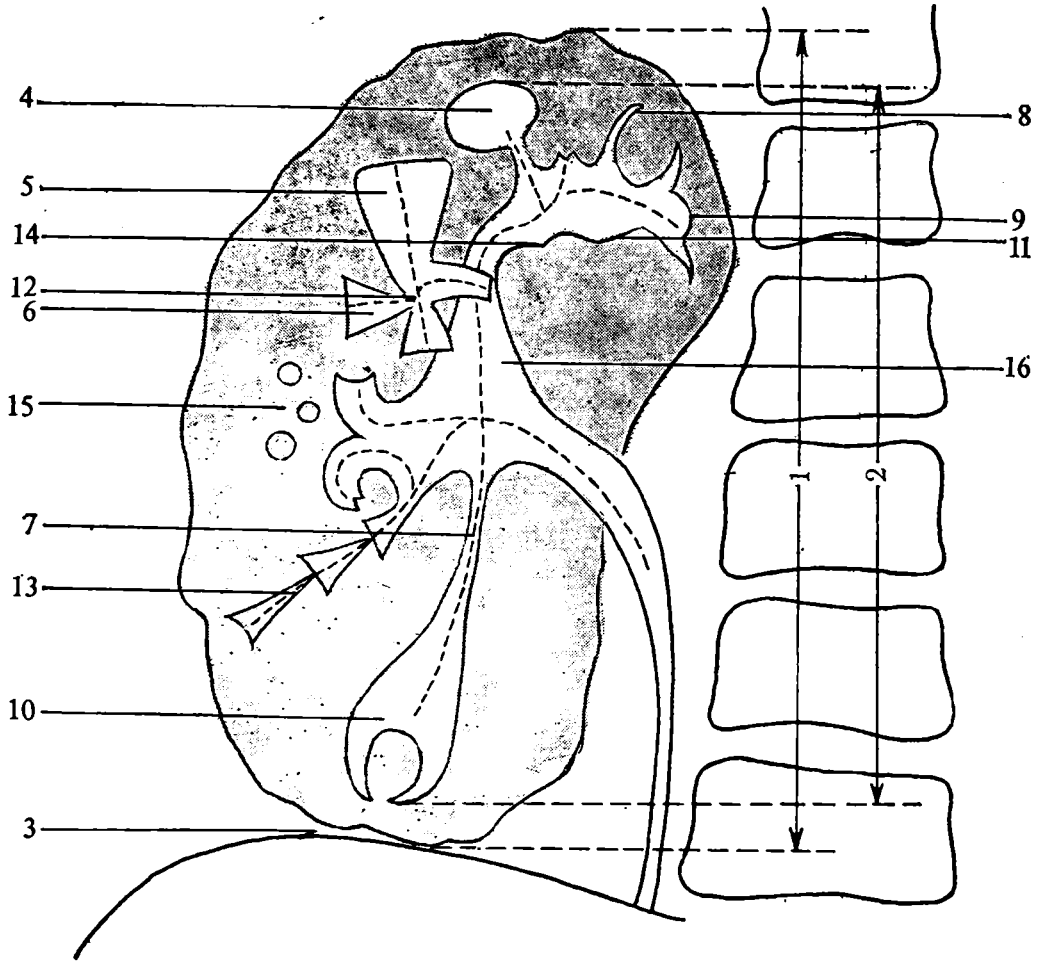


Fig. 26. Schematic representation of X-ray signs of polycystic kidney disease (after Hickel and Cornet, 1949).

1, increased distance between poles of the kidney; *2*, enlarged pelvic tree; *3*, downward displacement of lower renal pole; *4*, distended, spherical small calyces; *5*, petal-shaped, distended small calyces; *6,7*, elongated and narrowed neck of calyx; *8,9*, crescent-shaped image; *10*, lobster-claw image; 11, rosette symptom; *12*, calyceal axes arranged like wheel spokes; *13*, insertion symptom; *14*, crawling calyces; *15*, candy-type arrangement; *16*, cone-type image of superior calyx

Analysis of the ureteropyelogram pattern makes it possible to distinguish pathognomonic signs of polycystic kidney disease. They are subdivided by frequency into the most often encountered signs and auxiliary signs which are rare but very valuable for the diagnosis.

The main X-ray signs characteristic of the disease are as follows: enlarged total height of the renal shadow and the pelvic tree; abnormal position of the calyces; altered neck of the calyces; the initial part of the calyces is crescent-shaped or in the form of a spheric circumference; flask-shaped dilatation of the initial part of the calyces; increased number of calyces (hyper-

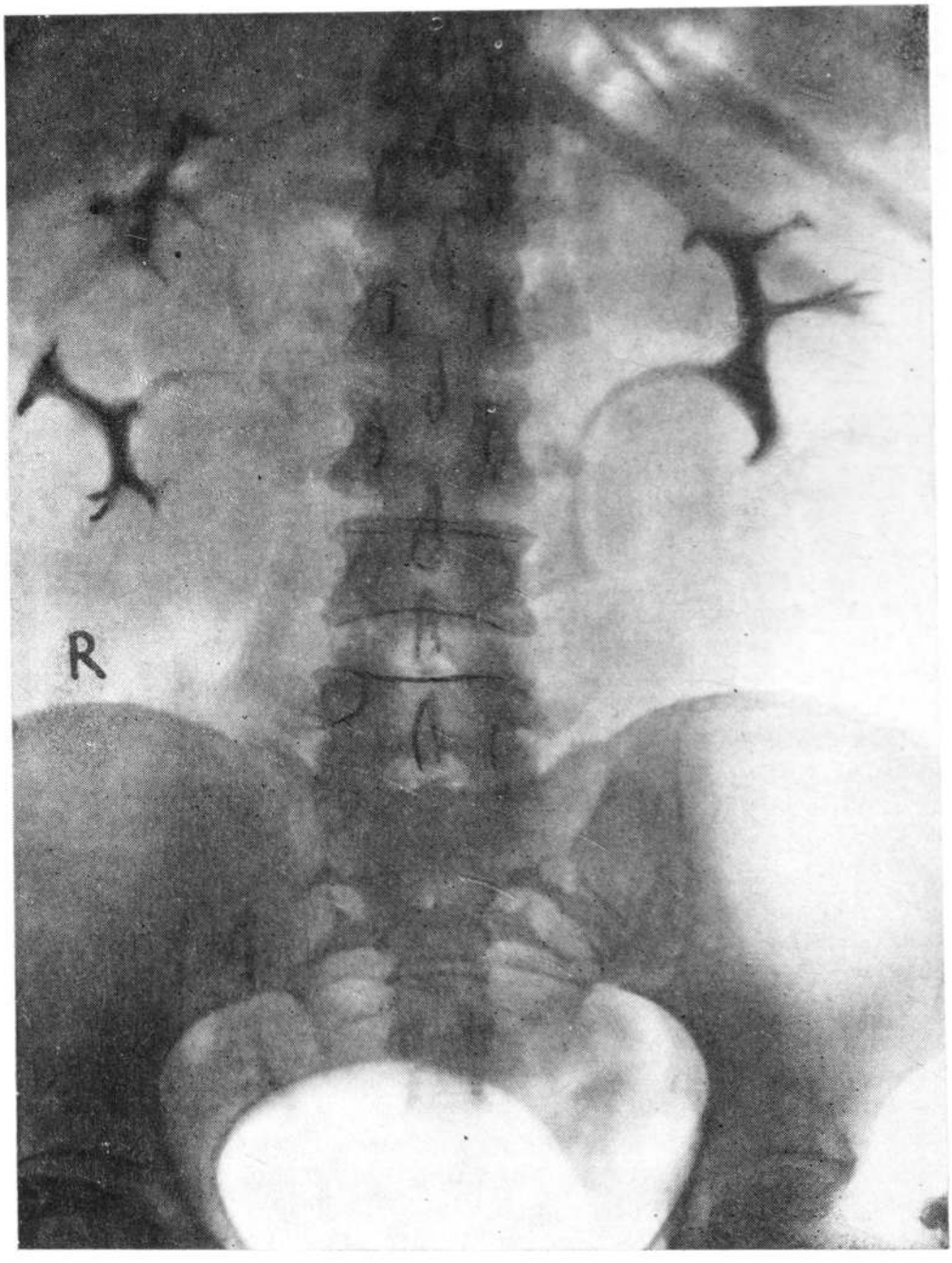


Fig. 27. Excretory urogram. Polycystic kidney disease, **duplication** of renal pelvis on the right

ramification); compressed and elongated pelvis; downward displacement of the pelvis.

The auxiliary X-ray signs which may prove valuable in making the proper diagnosis of polycystic kidneys are: diminished clarity

of the outlined image of the calyces; amputation or gross deformity of the calyces; distended and enlarged pelvis.

A schematical representation (after Hickel and Cornet 1949) of the X-ray signs of polycystic kidneys is given in Fig. 26.

Excretory urography. This functional and less traumatizing X-ray method of examination reflects the functional and morphological condition of the kidneys and upper urinary tract but, unfortunately, is valuable only in the initial stages of the disease, when renal function is still satisfactory.

Disorders of the functional capacity of the kidneys, manifested by a sharp decrease in concentrating ability and often encountered in polycystic disease, restrict the use of excretory urography. In the initial stage of the disease it yields very valuable X-ray data which, in combination with the general clinical findings, allow polycystic kidneys to be recognized without resorting to retrograde pyelography. There is no doubt that the development of new radiocontrast media (Cardiotrast, or diodone; Triiotrast, or sodium acetrizoate; Hypaque, or sodium diatrizoate) containing bi-, tri-, and tetratomic iodine promotes the wider use of excretory urography, especially its infusion form, in patients suffering from polycystic kidneys and, consequently, the procurement of results of higher diagnostic value.

When renal function is satisfactory, the shadow of the upper urinary tract on the excretory urogram is demonstrated just as clearly as in retrograde pyeloureterography (Fig. 27). This is achieved with the use of diodone (Cardiotrast) as the contrast medium and oxygen cystography suggested by Pytel (1953). It should be emphasized that before undertaking excretory urography in patients with polycystic kidneys it is very important to know the functional state of the kidneys and liver to avoid severe reactions and complications which may occur if their function is deficient. Patients may also have allergic reactions to the infusion of iodine-containing contrast media.

Thus, if the possibility of polycystic kidneys is borne in mind the disease can, in most cases, be diagnosed without much difficulty. Analysis of the patient's medical and family history is very important. Valuable information for making a diagnosis can be obtained from palpation of the kidneys. When it is found that both kidneys are enlarged and tuberous, then the diagnosis is clear. When only one kidney is palpable, a tumour or hydronephrosis should be suspected. It is very difficult to make a diagnosis when neither kidney can be palpated. Among the general clinical and laboratory methods of examination urinalysis, blood testing, tests for blood plasma urea and creatinine, and Zimnitsky's test prove valuable. The initial stage of polycystic kidney disease is characterized by mild proteinuria (up to 1 pro mille),

hyposthenuria, and increased ESR. With the development of inflammation pyuria, anaemia, and leucocytosis are observed. A history of hypertension indicates the need for scrupulous urological examination, not only for excluding unilateral renal lesion but also for recognizing polycystic kidney disease, bearing in mind that hypertension is encountered in 70 per cent of patients with this disease.

X-ray examination is essential in the diagnosis of polycystic kidneys. If the concentrating capacity of the kidneys is no less than 1.010 and there is no hyperazotaemia, X-ray examination should be begun with scout radiography and excretory urography. If the diagnosis remains unclear or doubtful after this examination, ureteropyelography is necessary.

We were able to recognize polycystic kidneys, often in the initial stage, in 82 per cent of patients by means of these methods in combination with general clinical examination. In doubtful cases where it was necessary to differentiate polycystic kidneys from tumour of the kidney or other cystic diseases (multicystic kidney, multilocular cyst, sponge kidney) or to diagnose a tumour in a polycystic kidney, we resorted to diagnostic pneumoretroperitoneum and renal angiography. We prefer the last method to other diagnostic techniques.

Complications. Polycystic kidney disease may be complicated by: (1) pyelonephritis; (2) urolithiasis; (3) new growth; (4) tuberculosis; (5) fornical haemorrhage; etc.

Pyelonephritis in polycystic kidney disease is a factor which indicates latent polycystic kidney disease and very often leads to the development of hypertension with severe renal insufficiency.

Among the symptoms making up the peculiar clinical picture of pyelonephritis in polycystic kidneys a protracted torpid course and bilateral affection take a special place. Dysplastic changes in the structure of the polycystic kidneys and disorders of blood circulation and often of the flow of urine in them are greatly conducive to this. It is they that cause the very protracted course of the disease and explain why it is difficult to eliminate the inflammation in the polycystic kidneys.

From analysis of our cases and the data in the literature, it should be pointed out that in the treatment of patients with polycystic kidneys complicated by calculous pyelonephritis the physician should remember that urolithiasis severely aggravates the main disease, therefore surgery is absolutely indicated in such cases. The earlier the stone is removed from the kidney or ureter, the less hazardous the possible complications will be.

A tumour is a rare occurrence in a polycystic kidney. However it is a menacing complication and the diagnosis of such a combined disease is very difficult. This is understandable since the

clinical manifestations of both diseases are similar in many respects. Renal angiography is indispensable in cases where diagnosis is particularly difficult.

Analysis of our observations and the data in the literature relating to pregnancy and polycystic kidney disease lead to the following conclusion.

There is no need to interrupt pregnancy if the disease is attended by subclinical disorder of renal function. In the initial stage of renal insufficiency, pregnancy sometimes terminates in normal delivery and the functional state of the kidneys does not grow worse. The question concerning pregnancy in this stage of renal insufficiency should nevertheless be approached comprehensively and in each individual case. The functional capacity of the kidneys often deteriorates sharply in pregnancy and abortion is necessary in such cases.

Pyelonephritis is a common complication in polycystic kidney disease, especially during a second or later pregnancy.

Pregnancy is life-endangering in women suffering from polycystic kidneys complicated by pyelonephritis and hypertension or attended by marked and severe renal insufficiency, and abortion is therefore recommended.

TYPES OF TREATMENT

Indications for non-operative treatment and surgery. Management of patients suffering from polycystic kidney disease and prolongation of their life are an important matter. The treatment of these patients is a difficult and complicated task, and since polycystic disease is a bilateral process which tends to progress, the results of the therapeutic measures are not radical or sufficiently effective.

The pessimism of physicians with regard to treatment and prognosis, however, is not altogether justified. Though the cystic renal tissue cannot regenerate and its function cannot be restored completely, appropriate regular treatment and examination and supervision of the patient may prolong his survival for many years. The main task is the timely prevention of complications, alleviation of the symptoms of the disease, and improvement of the functional state of the cystic kidneys and barrier systems and organs. It is known that the disease does not always develop to the stage of renal insufficiency; it often follows a latent course, the patients live to the age of 60 or 70 feeling quite healthy and retaining their capacity for work.

The therapeutic measures applied to polycystic kidneys may be distinguished according to the form of the disease as follows:

(1) for the uncomplicated form of polycystic kidney disease;

(2) for the complicated form of the disease;
(3) for polycystic kidneys with renal insufficiency.

People suffering from polycystic kidneys should be kept under constant surveillance with regular examination by specialists in different fields of medicine, and preventive and therapeutic measures should be continuously applied. All patients should be kept under observation by an urologist and therapeutist. Treatment will be successful only if what is begun in an in-patient hospital is continued in the out-patient clinic. Life dictates this condition. According to our data, in 86 per cent of cases polycystic kidneys are attended by pyelonephritis and hypertension, which call for long-term and regular treatment. Chronic pyelonephritis is one of the main factors inducing the change from the stage of compensation to that of decompensation. It also causes suppuration of the cysts as well as hypertension.

It is difficult to treat patients suffering from polycystic kidneys and various complications, among which pyelonephritis is most common. This group forms the main contingent of patients treated by urologists and surgeons, by therapeutists and neurologists, as well as by paediatricians.

Among the existing methods of therapy, we distinguish medicinal, dietetic, sanatorium, and surgical treatment.

We believe that surgery for polycystic kidneys is indicated in the following conditions: (1) persisting pain in the region of the kidneys; (2) large cysts, particularly in the region of the renal hilus; (3) recurrent total haematuria; (4) suppuration of the cysts; (5) hypertension which is resistant to hypotensive therapy; (6) tumour or tuberculosis of the polycystic kidney; (7) a stone in the kidney and ureter.

It is our experience that surgery (omentorevascularization) for polycystic kidneys will be successful only in the initial stage of renal insufficiency.

Surgery should not be performed in the subclinical stage of renal insufficiency because people with such a degree of impaired renal function may live for many years. Besides, rational non-operative treatment may delay the change from the subclinical stage to more severe renal insufficiency for many years. Surgery in the marked and severe stage (except for cases with suppuration of the cysts), which is recommended and undertaken by some authors as an extreme measure with the hope to improve renal function, is absolutely unjustified. The eight patients on whom we performed operation died soon after. Operative intervention in these stages of the disease does not prolong the patient's life but, on the contrary, shortens it. The question of surgery in the case of suppuration of cysts in the kidney is, of course, clear; an emergency operation for opening the cysts and inserting a

drain into them is necessary in such cases. Any type of surgical intervention performed on patients with polycystic kidneys is only one of the components of the treatment; after surgery all patients undergo long-term medicinal, dietetic, and sanatorium treatment.

Non-operative Methods

Non-operative management of polycystic kidney disease comprises a complex of measures: medicinal treatment and dietetic therapy combined with physiotherapy. Its purpose is to eliminate the infectious process in the urinary tract and control renal insufficiency, anaemia and hypertension.

Medicinal therapy is applied to most patients with polycystic kidneys mainly because they have pyelonephritis. Antibiotics and chemical agents are recommended. Antibiotics should be prescribed with caution because patients with polycystic kidneys usually have functional renal disorders and any antibiotic introduced into the body circulates in the blood for a lengthy period of time and when administered repeatedly may induce severe reactions and complications.

It should be noted that nitrofuranes (Furadonin *syn.* Nitrofurantoin, Furadantin) and Nitroxoline (also called 5-NOX, an abbreviation of 8-hydroxy-5-nitroquinoline) have a better effect than antibiotics. The efficacy of nitrofuranes is raised if the patients are given common horsetail decoction or cranberry extract per os before they take the drug. If nitrofuranes cannot be prescribed for some reason, the patient is given intravenous infusions of 5 ml of 40 per cent Urotropin (methenamine) solution for ten days and Nitroxoline or non-acetylating sulphanilamides per os. Medicinal therapy should be combined with an appropriate diet, physiotherapy and balneotherapy (pine baths, etc.).

Besides drugs and diet a complex of therapeutic measures for reducing azotaemia and eliminating nitrogenous residues from the body are prescribed at the stages of marked, severe, and terminal renal insufficiency.

It is our experience that patients with polycystic kidney disease complicated by pyelonephritis require long-term, regularly repeated medicinal and dietetic treatment. The prescribed treatment is aimed at the elimination or reduction of the inflammatory process in the urinary tract and improvement of the functioning of organs acting as barriers in the body, since this in turn prevents complications.

The prevention of pyelonephritis in patients with polycystic kidneys is a most important link in the treatment and in the prolongation of their life. This is a fundamental point which

should be taken into account in the regular surveillance of patients with polycystic kidney disease.

Surgical Methods

Because polycystic kidney disease is bilateral, great care should be exercised in deciding in favour of surgical treatment, nephrectomy in particular. At the end of the last century, the first operations for polycystic kidneys were usually of the nephrectomy type (Bobrov, 1891, and others). At that time most of them were undertaken for an erroneous diagnosis, usually for suspected tumour of the kidney. Waltern and Braasch (1934) reported that 16 per cent of patients died in the immediate period after nephrectomy, and 13.8 per cent died within the first 12 months after discharge from the clinic. Only two of 31 patients survived for 9 and 13 years after surgery. Prat (1960) is an advocate of nephrectomy for patients with polycystic kidneys complicated by pyelonephritis.

Analysis of the current literature and our experience, however, show quite convincingly that nephrectomy for polycystic kidneys is a hazardous operation and should be performed only in cases when cancer of the kidney is involved and rarely in cases of pyonephrosis. It is necessary to strictly abide by this rule because the immediate postnephrectomy mortality rate is very high and is mainly determined by the functional insufficiency of the remaining solitary cystic kidney. It is not surprising that the removal of one of the polycystic kidneys and, consequently, the reduction of the amount of functioning renal tissue in the body inflicts considerable operative trauma which is poorly tolerated by the patient. This is why patients die from various complications while still in the clinic or succumb to progressive renal insufficiency later.

Operation for opening, and emptying the cysts is performed very commonly.

In 1911 Rovsing suggested surgical decompression of a polycystic kidney by multiple punctures and emptying of the cysts. The operation is called ignipuncture. Eventually, many authors spoke in favour of this method, which was named Rovsing's operation. It is effective in the compensated stage of the disease. Puncturing and emptying the cysts reduce the size of the kidneys and intrarenal pressure and, therefore, improve circulation and the functioning of the nephrons. The operation improves renal function and relieves pain.

Some authors (Köhler, 1948; Ferrandu, 1953; and others) suggested a modification of Rovsing's operation involving the treatment of the cysts with sclerosing solutions after they had been

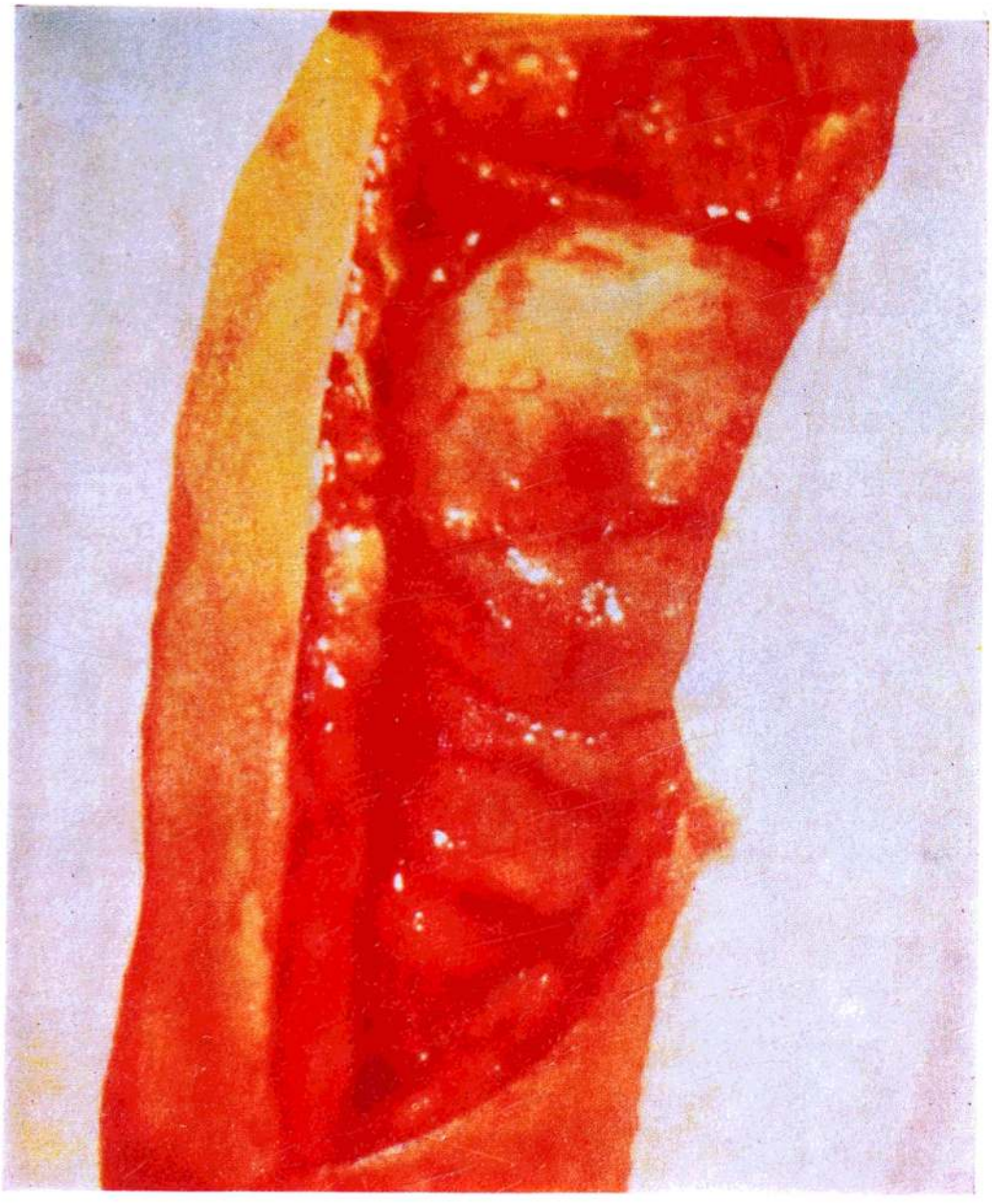


Plate I. Fedorov's oblique-transverse incision providing the best access to the kidney and abdominal organs. The peritoneum and retroperitoneal fat are seen deep in the wound

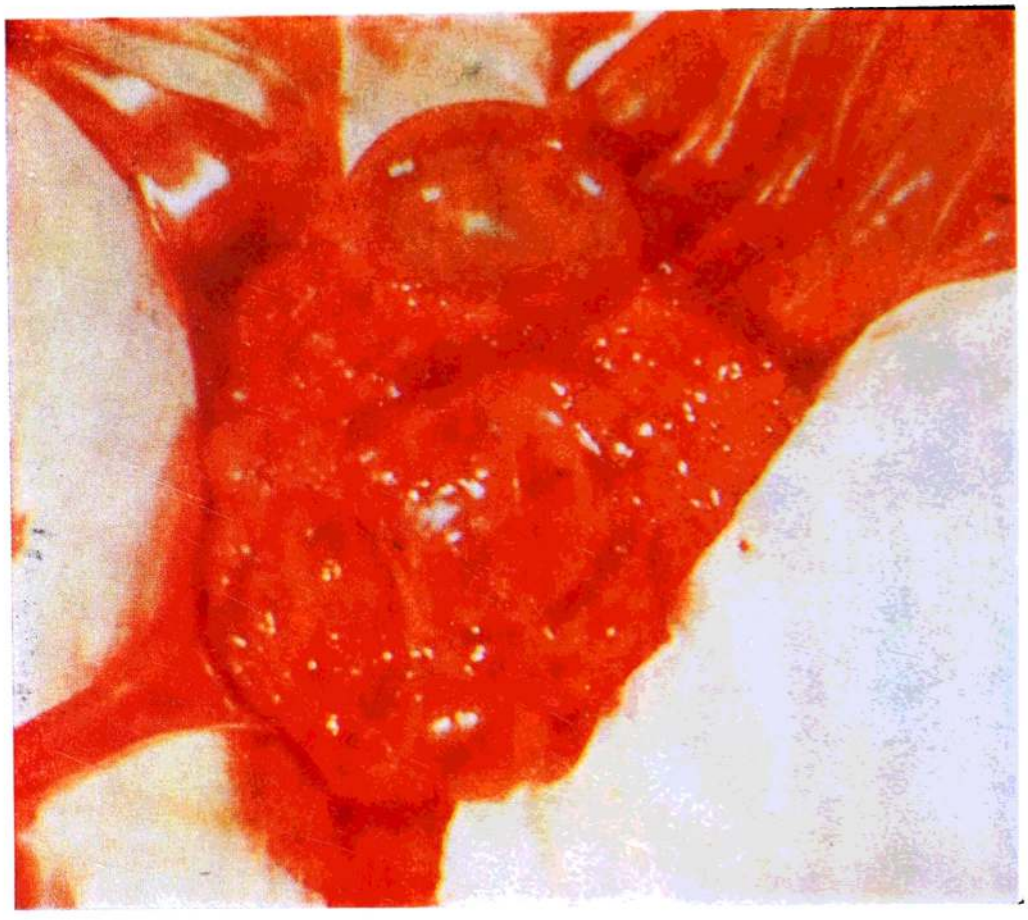


Plate II. The lower pole of the kidney is brought out into the wound; large and small cysts are seen on its surface; the region of the renal hilus is inspected

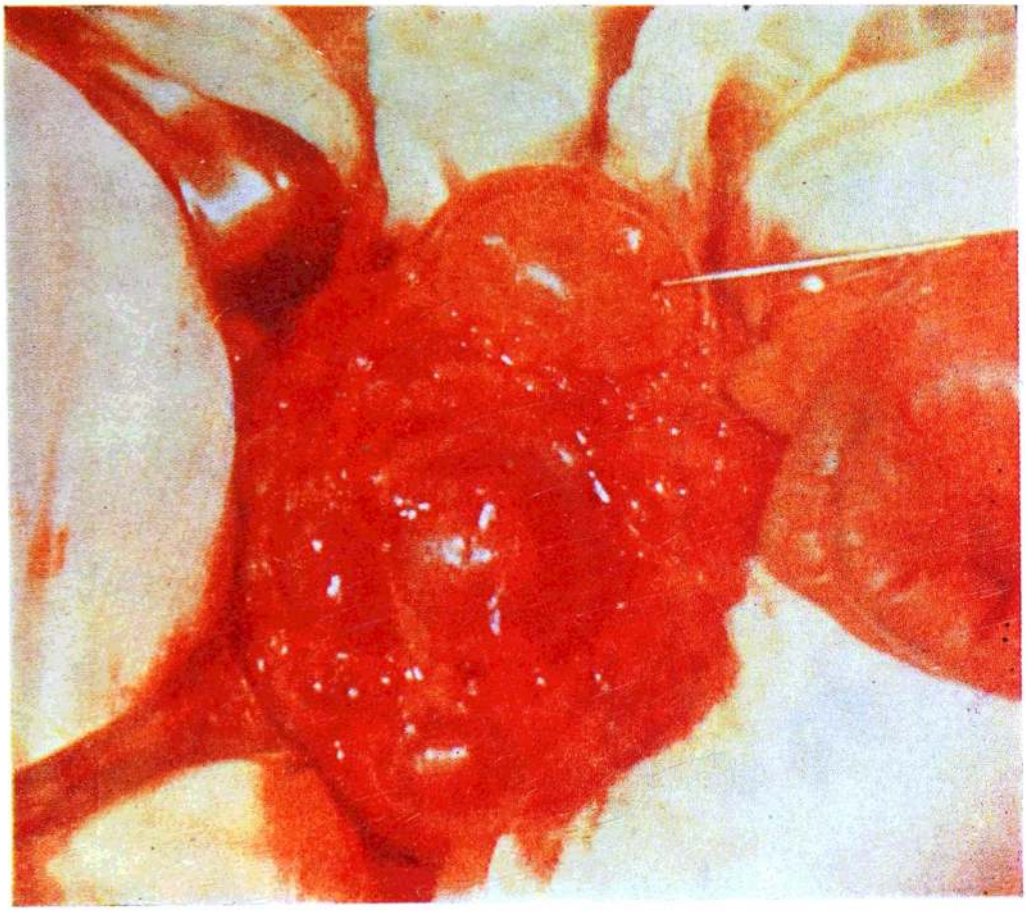


Plate III. Puncture of the large cysts and evacuation of their contents

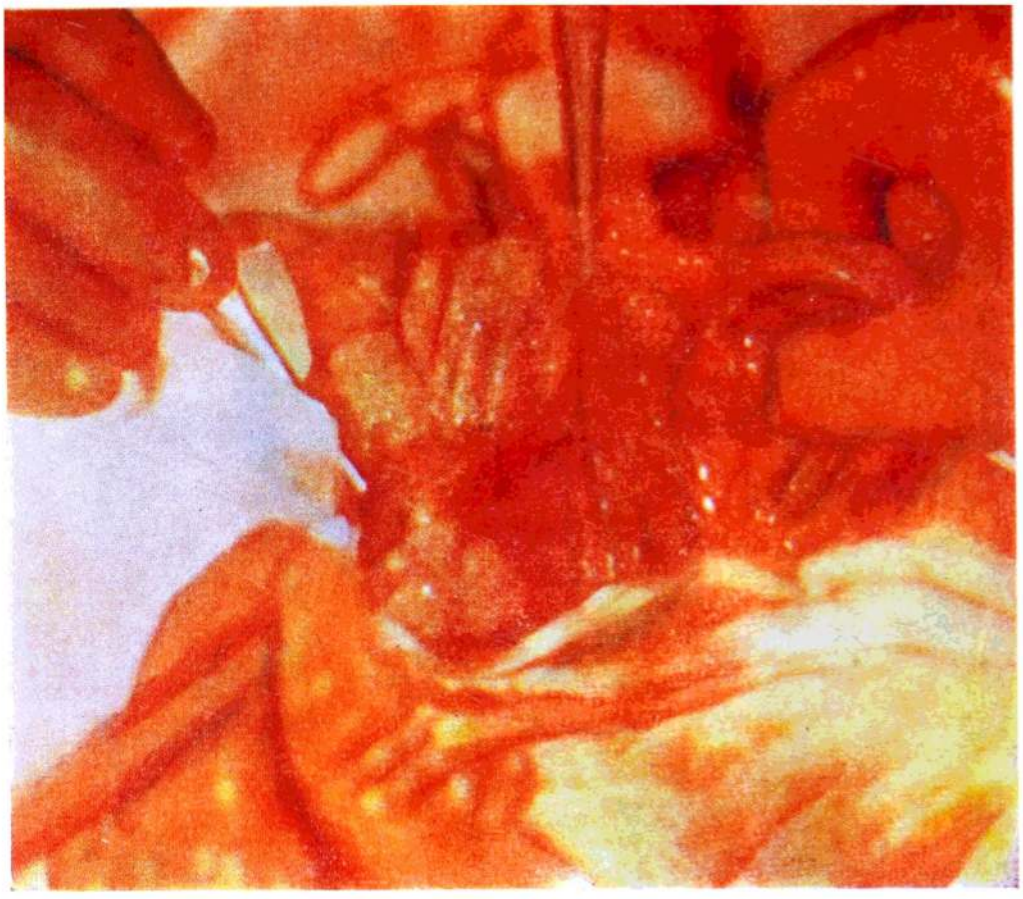


Plate IV. An opening is made in the peritoneum and the omentum is drawn out into the wound

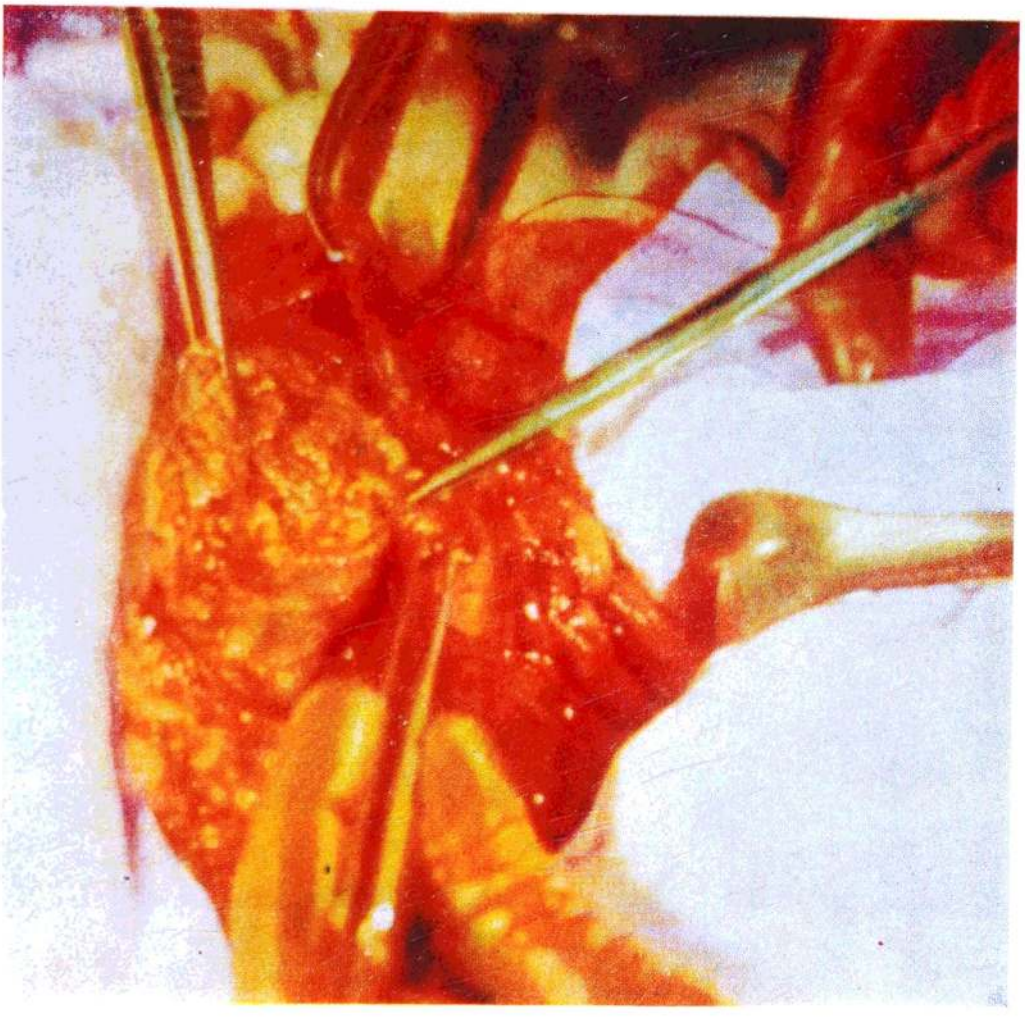


Plate V. The omentum is stitched to the edges of the peritoneal wound

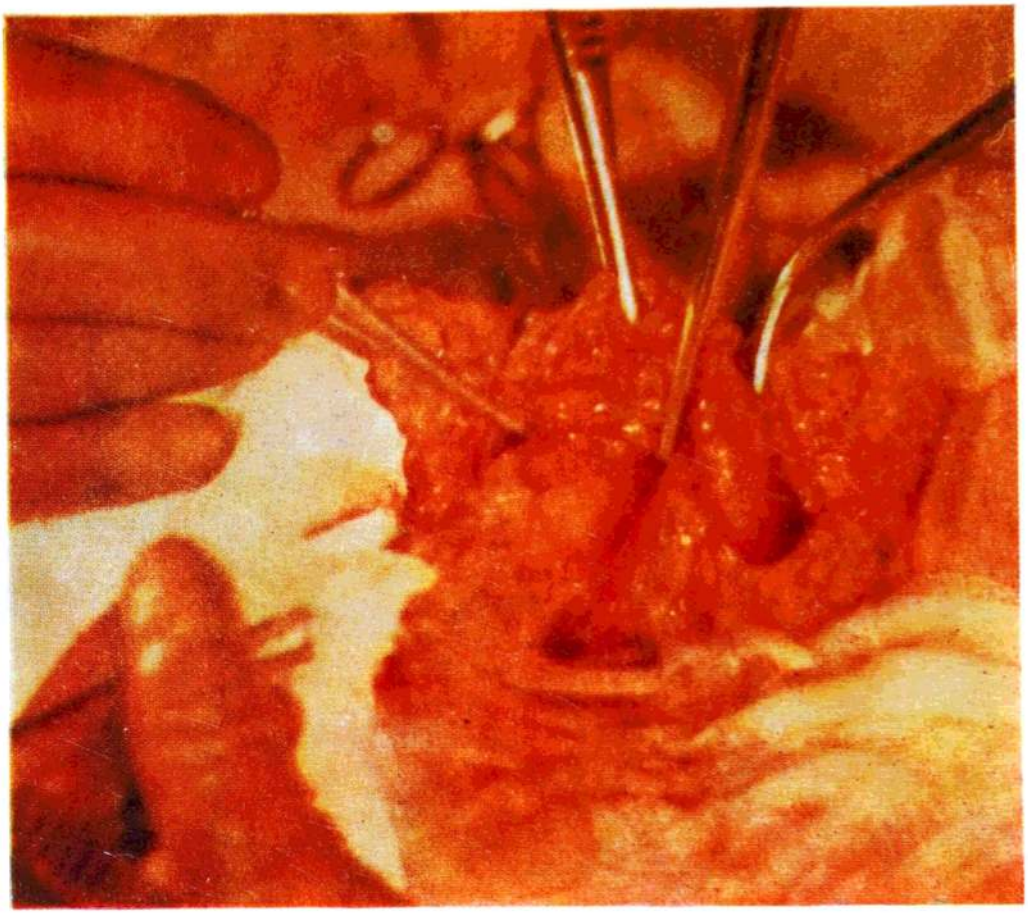


Plate VI. The omentum is drawn up to the kidney

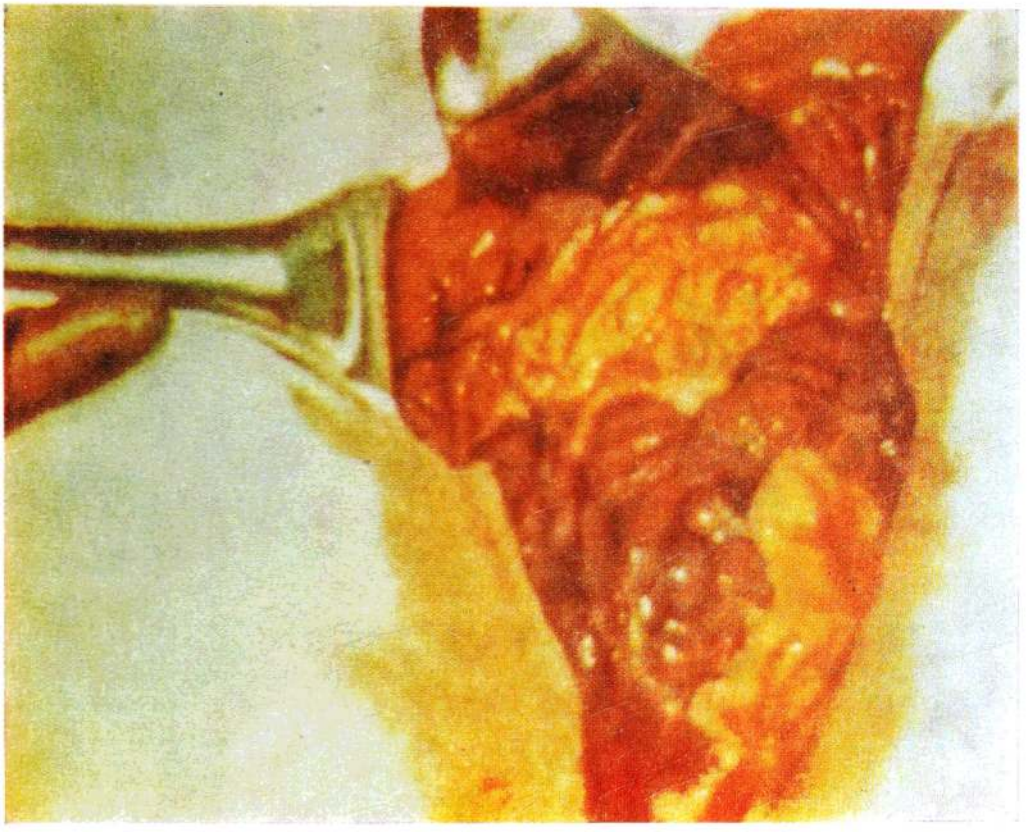


Plate VII. The omentum is stitched to the parenchyma on the anterior surface of the kidney. At the bottom, perirenal fat is packed into the cavity of the large cyst and fastened with sutures

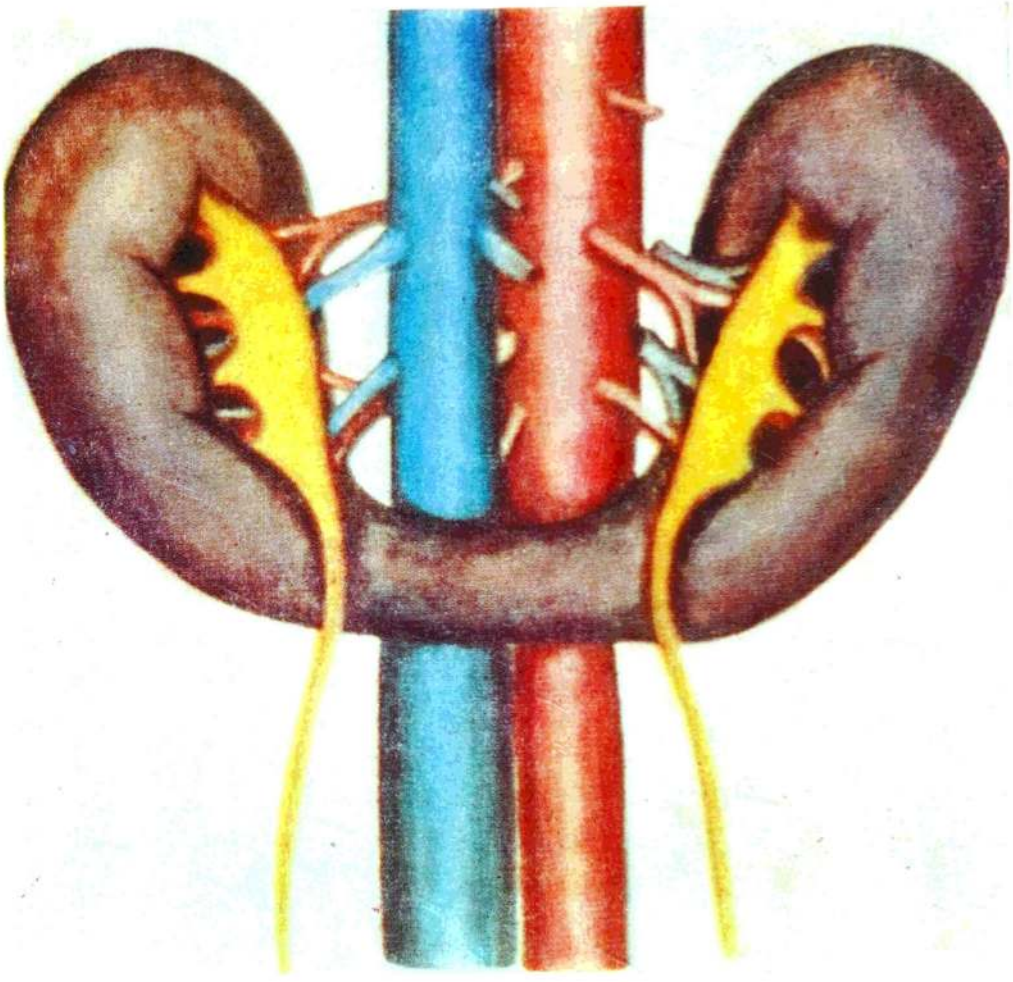


Plate VIII. Compression of the aorta and vena cava inferior by the isthmu of a horseshoe kidney

opened and emptied. For this purpose, Young (1946) used 5 per cent sodium chloride solution. After emptying the cysts, Ferrandu introduced into their cavity equal parts of 1.5 per cent monothanolamine emulsion and paraffin oil; a good clinical effect was produced in three patients (follow-up periods of two to seven years). These methods, however, did not gain supporters. In 1935 Goldstein (1951) suggested a surgical method comprising transplantation of the polycystic kidney under the skin after the cysts had been emptied and their walls excised. Fixation of the kidney under the skin allows punctures to be repeated.

Another operation used in the treatment of patients with polycystic kidneys, percutaneous puncture of the cysts, deserves to be noted. It was suggested by De la Pena (1961).

Of particular interest are the results of the use of an artificial kidney in patients with polycystic kidney disease complicated by renal insufficiency. In reference to this, the work of Nakamoto and Kolff (1958) is worthy of notice. It is the first report on the treatment of 13 patients with polycystic kidney disease complicated by chronic uraemia.

We shall dwell here briefly on kidney homotransplantation in the treatment of patients with polycystic kidneys. Judging from the data in the literature, successful transplantation of a homologous kidney has been performed in six patients. Transplantation of a kidney from a monozygotic twin has been carried out dozens of times. In 1952, Lawler transplanted a human kidney into a 44-year-old woman with polycystic kidney disease with due consideration for the identity of the blood group and sex of the donor and recipient. The donor was a 49-year-old woman who had died from cirrhosis of the liver. The nearest relatives of the recipient also had polycystic kidneys.

A very interesting case was described in the literature, with transplantation of a kidney from a brother to a sister (they were not twins) who had polycystic disease and uraemia (Woodruff et al., 1962). Unfortunately, in both cases transplantation of the kidney was unsuccessful.

Thus, in the treatment of patients suffering from polycystic kidney disease and attendant renal insufficiency a question always arises concerning the application of substitution therapy, namely haemodialysis and kidney transplantation. Our experience shows that in individual cases the course of the disease and the functional condition of the kidneys improve during haemodialysis to such a measure that removal of the arteriovenous shunt and application of non-operative treatment may be contemplated. We encountered this in three patients, the remission in these cases lasting eight months to two years. Sometimes, in the severe and terminal stage of renal insufficiency it proves

possible to improve the patient's condition after three or five sessions of haemodialysis, to reduce the high arterial pressure, and enable him to resume work; depending on the circumstances and place of work haemodialysis is conducted under out-patient clinic conditions or at night. The task is more difficult in the decompensation phase of the terminal stage when the functional capacity of the kidneys and other organs (heart, liver, lungs) is exhausted. True, haemodialysis has an amazing effect at this stage in some cases, the patient recovering from this critical condition and may be prepared for kidney transplantation. Usually, however, the patient's life is prolonged only by six to ten months. The patients die from cardiovascular failure and concomitant infection.

The results of kidney transplantation are less favourable in polycystic kidney disease than in other morbid conditions. This is evidently explained by the possibility of the associated incidence of cystic lesions in other organs, especially the liver. During haemodialysis conducted in preparation for kidney transplantation the possibility of the removal of the cystic kidneys has always to be considered. We prefer removing them in two stages: one before the donor kidney is transplanted, and the other during transplantation. Patients tolerate one-stage binephrectomy poorly and its outcome is often fatal.

These principles of the treatment of patients with polycystic kidneys involve all current advances in uronephrology and prolong the life of patients with this grave disease by many months or even years.

Analysis of the surgical treatment of our patients shows that ignipuncture was the most commonly used manipulation. Since 1959 this operation, opening and emptying of the cysts, has been performed in combination with omentorevascularization of the kidney by a method developed at our clinic. Our method employs general anaesthesia (nitrous oxide inhalation anaesthesia with intubation and the use of muscle relaxants). Federov's lumbar oblique-transverse incision provides good access to the kidney and abdominal organs (Plate I). The kidney is exposed and inspected, particularly the region of the hilus, for large cysts (Plate II). The contents of the large cysts are then aspirated and their walls excised (Plate III). It is not recommended that small cysts of up to 1.0-1.5 cm in diameter be destroyed. The abdominal cavity is opened. The liver is examined, small cysts are sometimes found on its surface; these should not be emptied, but the contents of large cysts with a diameter of more than 3 cm are aspirated. The greater omentum is inspected and divided in half approximately at the level of the middle of the transverse colon; one half is then mobilized up to the splenic or hepatic

flexure (depending on the side of the intervention), and brought out easily through an additional opening in the parietal peritoneum and drawn up to the kidney (Plate IV). The opening in the parietal peritoneum is closed by stitching the omentum to its edges (Plate V). The abdominal cavity is sutured completely. An incision 6 to 8 cm long and 2 to 3 cm deep is made on the surface of the kidney and a small piece of the parenchyma is removed for histological examination to determine the depth of the inflammatory process in this kidney. The omentum is fitted into this wound in the kidney and sutured with interrupted catgut sutures (Plate VI). Careful haemostasis is performed. Perirenal fat is packed as a tamponade into the cavities of the large cysts with resected walls (Plate VII). A gauze-rubber tampon or a silicone tube is advanced to the lower pole of the kidney and left there for four or five days, with sutures being applied to each layer separately.

We recommend performing the second operation on the other kidney after 12 to 18 months as a rule.

We performed operations by this method on 145 patients; in one omentorevascularization was combined with resection of the upper pole of the kidney.

Clinical examination and laboratory tests conducted in the postoperative period showed that the filtration-reabsorption ability of the kidney operated on by this method had improved.

It should be noted that operating for the omentorevascularization of a polycystic kidney combined with ignipuncture leads to a reduction of intrarenal pressure and, consequently, to a decrease in venous and lymph stasis. Besides, it is obvious that evacuation of the contents from the large cysts (up to 600 to 800 ml), decompression of the cysts, and stitching of the omentum, rich in blood and lymph vessels, into the parenchyma of the cystic kidney improve the circulation of blood and lymph in it. The possibility is created for the formation of vascular anastomoses between the cortex and the omentum. Hypertension was relieved in 83 of our patients after the operation.

As for operations in cases of polycystic kidneys and associated renal insufficiency, it should be emphasized again that such patients are exposed to great danger because of impaired homeostasis and complications in the cardiovascular system, lungs, liver and other organs. However, there are sometimes vital indications for carrying out the operation even in cases of marked and severe renal insufficiency. Suppuration of the cysts and profuse haematuria may make surgery necessary in a patient who is in such a grave condition. In our practice, three patients with marked renal insufficiency underwent ignipuncture of cysts which

had caused attacks of acute pain in the renal region with exacerbation of the inflammatory process in the kidneys.

To reduce the risk of surgery in this category of patients, we now use active methods of preoperative treatment (Dzhavadzade and Malkov, 1974). In cases of marked and severe renal insufficiency non-operative therapeutic measures usually cannot correct the impaired homeostasis, relieve uraemic intoxication or alleviate the severity of the hypertension syndrome and clinical course of the disease.

A therapeutic effect may be produced in preoperative management if haemodialysis is included in the complex of measures. Extracorporeal haemodialysis brings rapid relief from uraemic intoxication, normalizes homeostasis and improves the clinical course of the disease, which reduces the risk of the operation.

The operative trauma itself may sometimes exacerbate renal insufficiency with intensification of hyperazotaemia and uraemic intoxication. The application of haemodialysis as therapeutic insurance may play a decisive role in the outcome of the operation and in the postoperative period.

In two cases we observed aggravation of initial renal insufficiency after omentorevascularization, which called for three to seven sessions of haemodialysis.

Anaesthesia also presents a certain risk in renal insufficiency. In this group of patients, it is no less hazardous than the operation itself. This is why neuroleptanaesthesia with Droperidol (dehydrobenzperidol) and Fentanyl (phentanyl, R-4263) is the method of choice.

The data in the literature and our experience have convinced us that these agents possess only mild toxicity in relation to liver and kidney function. They reliably protect the body from the effect of stress factors and contribute to the favourable course of the postoperative period.

Prognosis. It is our experience that in most cases the disease follows a favourable course if complications occur in a patient over 40 years of age. The prognosis is very poor when complications develop in individuals under the age of 30 to 40. This circumstance brings problems of early diagnosis even more to the forefront. In view of this, besides the general clinical methods of examination the newest laboratory tests and radioisotope and X-ray techniques should be widely used for the recognition of the disease in doubtful cases. Early diagnosis of the disease, timely therapy and regular medical surveillance of patients are the principal means of prolonging the life and working capacity of people suffering from polycystic disease.

CHAPTER III

Fused Kidneys

(ANOMALIES IN SHAPE AND INTERRELATION OF KIDNEYS)

Fused kidneys have been known since the 16th century. According to Beyer (1912), Berenger de Carpi was the first to publish in 1522 information about a horseshoe kidney. Since that time, three periods in the study of fused kidneys should be noted: (1) the period of autopsy diagnosis, which continued to the second half of the 19th century; (2) the period of operative diagnosis, when the first attempts at clinical diagnosis and surgical treatment were made; it lasted into the first decades of the 20th century; (3) the period of preoperative diagnosis, scientific study of the clinical course of the anomaly and its diseases, and the peculiarities of surgical management. Some problems concerning the diagnosis of the anomaly and different methods of surgical treatment are being elaborated particularly actively in recent years.

Martynov, Fedorov, Gotlib, and many other Russian and then Soviet urologists focused much attention on the study of this anomaly in their work. In the Soviet literature, fused kidneys are discussed in Gimpelson's monograph (1949) and by Shimkus in his dissertation (1969). The first work is based on the data of 35 cases, the second contains information on 111 clinical cases and 12 postmortem examinations. Besides these there are also many publications based on a small number of observations.

In other countries, the problem is discussed in Gutierrez' monograph, published in 1934 and based on the study of 25 clinical cases and postmortem material, and in Pepersack's (1961) sur-

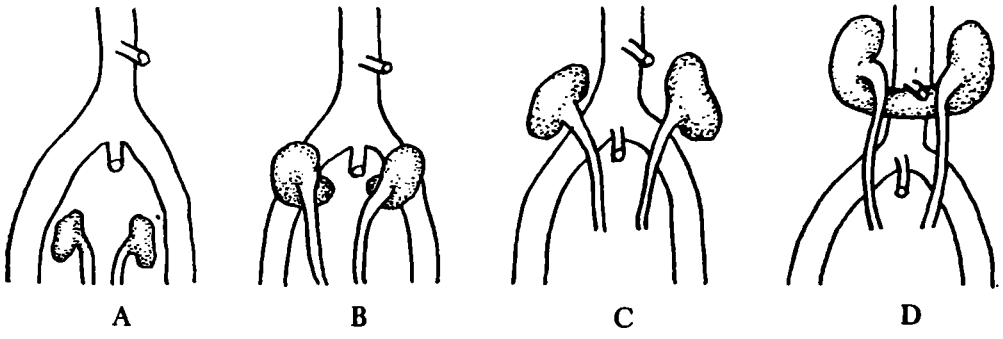


Fig. 28. Diagram showing the migration of kidneys from pelvis.
A, in pelvis; *B*, between umbilical arteries, where lower poles may fuse; *C*, kidneys free to migrate upward; *D*, horseshoe form caught under inferior mesenteric artery (after L. Bremer, 1957)

vey of the literature on horseshoe kidneys. All other works are articles published in journals, most of which are based on individual observations. Exceptions among these are reports made by Zondek and Zondek (1964) on a rich postmortem material (99 cases) and by Culp and Winterringer (1955) who described 106 cases observed in the clinic.

Fused kidneys evidently form in the eighth week of embryonal life during the formation of the permanent organs. Bremer (1957) gives an interesting explanation of the pathogenesis of this anomaly in his work dealing with research into the development of various human anomalies. He believes that in migrating cranially the kidneys have to push through a crotch formed by the two umbilical arteries, the end branches of the abdominal aorta. This brings the lower poles of the kidneys close together. The unfavourable effect of different factors at this time can keep the kidneys in that position, while intensive cell proliferation attendant with embryonal development can promote fusion (Fig. 28).

The fusion of kidneys before their complete ascent and rotation preserves the pelves on the anterior surface of the organ.

The mesenteric vessels prevent complete cranial migration of the fused kidney as a result of which a Y-shaped curvature and an isthmus are formed. Kidneys thus shaped always remain at the middle lumbar level and their further migration is restricted by the root of the inferior mesenteric artery. A fibrous isthmus may evidently sometimes form from stronger pressure against the root of this artery. In this case, the kidney may migrate higher, almost to the normal level. This explains symmetric fusion of the kidneys at the lower poles. But with the predominance of one half of the kidney, the other half moves after it and the migration is continued, by-passing the root of the inferior mesenteric artery. In this case asymmetrical forms of fused kid-

neys are formed (angulated and unilateral fused kidneys situated to one side of the spine, and their numerous varieties).

To explain the formation of horseshoe kidneys fused at the upper poles, and cake and sigmoid kidneys it has to be assumed that during migration of the kidneys conditions may be created under which the organs come together at the upper poles or the middle parts, or the upper pole of one kidney fuses with the lower pole of the other.

Smirnov (1959) offers another explanation of the genesis of this anomaly. He suggests that, at the time that the protrusion of the Wolffian duct grows into the metanephrogenic tissue, this tissue separates into two parts. But if the ureteral buds approach the metanephrogenic tissue before it has separated, perhaps we can assume that the delay leads to partial or total fusion of the kidneys.

CLASSIFICATION

Fused kidney is a term describing a congenital anomaly characterized by the fusion of two kidneys into a single organ whose ureters drain into the urinary bladder at the usual sites. The embryological data permit us to assume that fused kidneys differ in the type and extent of the fusion and in the position and shape of the organ. The final position and shape of such a kidney depend on the type of fusion.

The difference in the extent of the fusion, the size of the fused parts, the position in relation to the spine, and other anatomical features determine the large variety in the external shape of the fused kidneys. Each of these numerous varieties has a name of its own. Fusion of the upper or lower poles of two kidneys on the midline of the body in the form of a horseshoe is called a horseshoe kidney. This anomaly is also known as arched, hoof-like (ungulate), crescent, or unpaired kidney (Bukeikhanov, 1941). When one kidney is located normally, while the other is fused with it to form a right angle and lies across the spine, the condition is called angulated, L-shaped, or hooked kidney (Gotlib, 1938; Parin, 1938). If the fused kidneys are one below the other on one side of the spine and their hilus face the same direction, such a kidney is called unilateral fused kidney (Fronshtein, 1929), unpaired fused (Resh, 1945), double unilateral fused (Baranov, 1925), double fused (Farkash and Revestz, 1930), double solitary (Ivankovich, 1926; Chaika, 1929), elongated (Kreuzbauer, 1927), or crossed dystopic fused kidney (Lus, 1954). A fused kidney located to one side of the spine with the hilus facing in different directions is known as sigmoid, or S-shaped kidney (Brosch, 1957). Kidneys fused at the lateral edges or the middle parts

are called cake, double fused pie, shield-shaped, nail-shaped, doughnut, disc, block-like, clump, or shapeless kidney (Lavrov, 1928; Schmid, 1966).

The great number of varieties of shape and the use of more than one name for each of them or, sometimes, the use of one and the same name for the different types have introduced complexity and confusion into the terminology. That is why even a solitary double kidney with a bifurcate ureter has sometimes been taken for a fused kidney (Yurgelevich, 1922). The literature contains descriptions of an angulated kidney with duplicated urinary tract which is erroneously called a sigmoid kidney when there is a third, supernumerary kidney (Markin, 1960), or of a horseshoe kidney with duplication of the urinary tract which is called "crossed dystopia of a sigmoid kidney" (Balueva, 1964).

The terminology is also complicated because the classification of fused kidneys has its shortcomings. In some classifications the anomaly is considered from the standpoint of embryogenesis. Zakharyan (1941), for instance, proceeds from the assumption that the low position of the kidney is the main symptom of the anomaly, while fusion is secondary and not obligatory. That is why he includes fusion of the kidneys in the group of dystopias. Other classifications (Bogdansky, 1950) take into greater consideration the anatomical signs. In this case the different variants of the structure of the renal tissue and upper urinary tract are presented in detail, and combinations of the anomaly with other developmental anomalies of the kidneys are distinguished. Classifications drawn up according to embryological or anatomical sign are inconvenient and difficult to use in the clinic because some do not include the main anatomical types which differ considerably one from another, while others are too detailed and contain numerous varieties of no great importance. These classifications are of interest to embryologists and morphologists but not to clinicians.

In older clinical classifications fused kidney was included among abnormalities of number together with kidney aplasia (Bugbee and Losee, 1919), abnormalities of shape (Arrigoni and Vaccheri, 1962) or position (Zakharyan, 1941), or anomalous interrelations of the kidneys (Gimpelson, 1949). It should be noted that in these classifications the same types of abnormalities can be often included in different groups. For example, Smirnov (1958) places horseshoe kidney in the group of anomalous interrelations, and homolateral dystopia with fusion in the group of renal dystopies. Arrigoni and Vaccheri (1962) describe unilateral fused kidney as being in the group of anomalous interrelations, and crossed dystopia of kidneys with fusion among renal dystopias. To this day, unilateral fused kidneys are related to anomalous interrela-

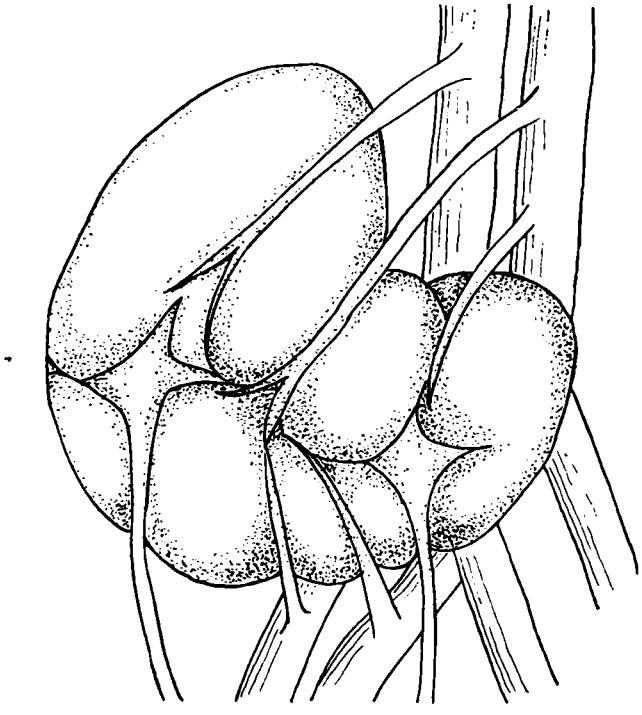


Fig. 29. Cake kidney of D., situated to the right of the midline (drawing made during operation)

tions in some classifications (Marton, 1958) and to abnormalities of position in others (Gimpelson, 1949).

Gimpelson's classification is based on the determination of the shape and position of the organ. All fused kidneys are separated into two groups, symmetric and asymmetric, depending on the position in relation to the midline of the body. Each group in turn is subdivided respectively into horseshoe and cake kidneys, and L-shaped and sigmoid kidneys. This classification, however, also has serious shortcomings. The strict division of all anatomical types of fused kidneys into symmetric and asymmetric forms cannot but give rise to doubt. It is not only that the concept of symmetry itself as applied to this anomaly is very relative, but that the literature contains descriptions of symmetric variants of types which, according to this classification, should only be asymmetrical, for instance, as is the case with a symmetrical sigmoid kidney. We encountered an asymmetrical cake kidney (Fig. 29), which has no definite place in Gimpelson's classification.

The modern concepts of the embryogenesis of fused kidneys do not justify the exclusion of unilateral fused kidney from the group of anomalous relationships: it shares a common mechanism

of development with the angulated and horseshoe kidneys fused at the lower poles.

The way the kidneys are fused affects their anatomical and clinical features. That is why it is undesirable to classify all horseshoe kidneys fused at the upper and lower poles in one group, and kidneys fused at the middle in the group of cake kidneys.

Gimpelson's classification does not allow for intermediate forms between some of the anatomical types, for example between angulated and sigmoid kidneys, which are classified next to each other in one group but differ in the type of fusion. At the same time, anatomical types related in embryogenesis, namely horseshoe kidneys fused at the lower poles and angulated kidneys, are classified in different groups. This makes it difficult to reveal the connections between all the main types of fused kidneys and does not allow one to form a complete idea of the whole group of anomalous relationships of the kidneys.

The numerous anatomical varieties of fused kidneys described in the literature show that there is a connection between the main anatomical types. Intermediate forms are easily found between a horseshoe kidney fused at the lower poles, and angulated and unilateral fused kidneys (Fig. 30).

It is likewise easy to find intermediate forms between horseshoe kidneys fused at the upper poles and sigmoid kidneys (Fig. 31). One can also conceive varieties intermediate between horseshoe and cake kidneys. This confirms once more that these anomalies share a common genesis and that it is desirable to classify them in one group.

We believe that the main shortcoming of all the suggested classifications consists in the fact that taking into account the shape and position of the anomalous organ, they do not give due consideration to the way in which fusion has occurred, and

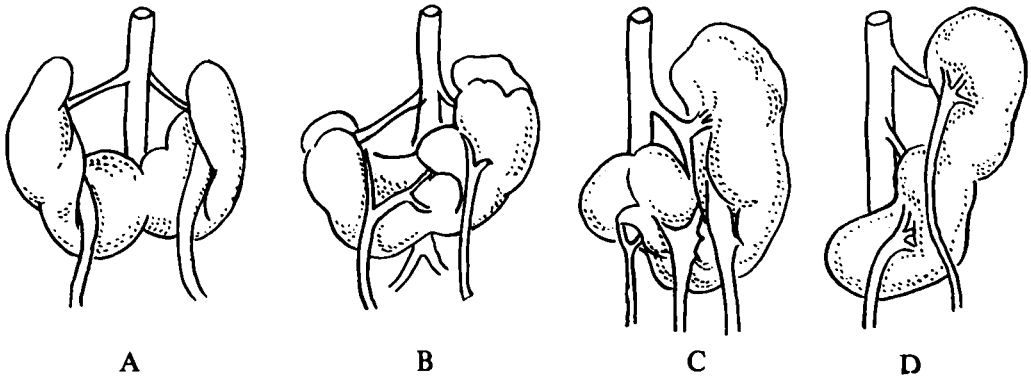


Fig. 30. Some varieties of anomalies in fusion of kidneys at the lower pole. *A*, *B*, horseshoe kidney; *C*, angulated kidney; *D*, unilateral fused kidney

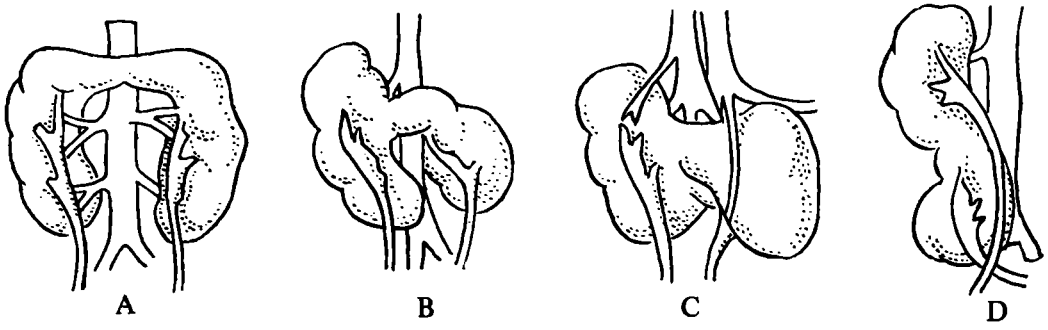
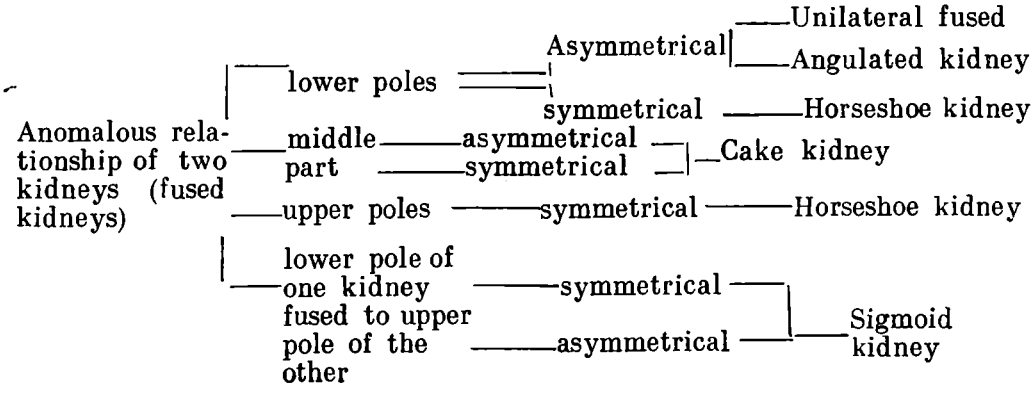


Fig. 31. Varieties of anomalies in fusion of kidneys at the upper poles or in fusion of the lower pole of one kidney with the upper pole of the other.
A, horseshoe kidney; *B*, fusion of the upper pole of the left kidney with the lateral surface of the right kidney (after Gutierrez, 1931); *C*, fusion of the upper pole of the left kidney with the lower pole of the right kidney (after Anichkov, 1912); *D*, sigmoid kidney

it is this which determines the final shape and position of the fused organ. To avoid this, in our classification we take into account the following three signs: the way of fusion, the position and the shape of the kidney. The position of the urinary tract and the renal vessels is of minor importance in these cases and we therefore disregard it.

We separated all fused kidneys encountered by us into four groups according to the way in which they are joined: at the lower poles, in the middle parts, at the upper poles, and a group in which the lower pole of one kidney is fused with the upper pole of the other. Each group in turn is subdivided according to the position of the kidneys into relatively symmetrical and asymmetrical forms and then, according to the shape of the organ, into six main anatomical types: horseshoe kidneys fused at the lower poles, angulated, unilateral fused, cake kidneys, horseshoe kidneys fused at the upper poles, and sigmoid kidneys. Thus, the extreme types in the group of fused kidneys are unilateral fused and sigmoid kidneys, between which all the other main anatomical types and their varieties can be easily placed as follows.



This classification is in compliance with the embryological data and presents the entire group of fused kidneys. It enables one to distinguish all the main anatomical types and determine the place in it of all possible varieties. The classification makes it possible to present in a single group the embryogenetically related unilateral fused and angulated kidneys and horseshoe kidneys fused at the lower poles; to determine easily the type to which any variety belongs; to find the place for the asymmetrical variants of cake kidneys and symmetrical variants of sigmoid kidneys described in the literature or encountered in our practice; and to allow that horseshoe kidneys fused at the upper poles also have asymmetrical variants. We consider this classification simple and convenient for use in clinical practice.

SURGICAL ANATOMY

The difference in the extent and way of fusion of the kidneys as well as in their shape and position in relation to the spine creates a great number of intermediate varieties of fused kidneys. Therefore one fused kidney never resembles another in its details and thus cannot be given a strictly definite description as in normal anatomy. Nevertheless, the different anatomical variants of this anomaly have much in common.

MORPHOLOGICAL STRUCTURE

The **renal parenchyma** of the fused organs is characterized by its diverse shape. We encountered different variants of fused kidneys, some of them are shown in Figs. 32-34.

When fused at the poles, the kidneys have the appearance of a horseshoe with the concavity facing upward or downward. We encountered fusion of the lower poles in 203 cases and fusion of the upper poles only in three.

According to Carlier and Gerard (1912) and Pepersack (1961), fusion of the kidneys at the lower poles is encountered in 90 to 95 per cent of cases. Dorland (1911) described a case in which the poles of a horseshoe kidney faced the abdominal cavity.

Angulated, unilateral fused and sigmoid kidneys are more elongated or bent. Cake kidneys are particularly quaintly shaped. They may resemble a pie or nail. Garbarets (1966) described a kidney fused at the middle parts and resembling a butterfly. Among the variants of cake kidney, the block-like or clump kidney deviates most in shape. It is composed of a shapeless mass of renal tissue divided by deep grooves.

Kidneys often fuse by means of an isthmus. The common character of the renal tissue and vascularization suggests that the

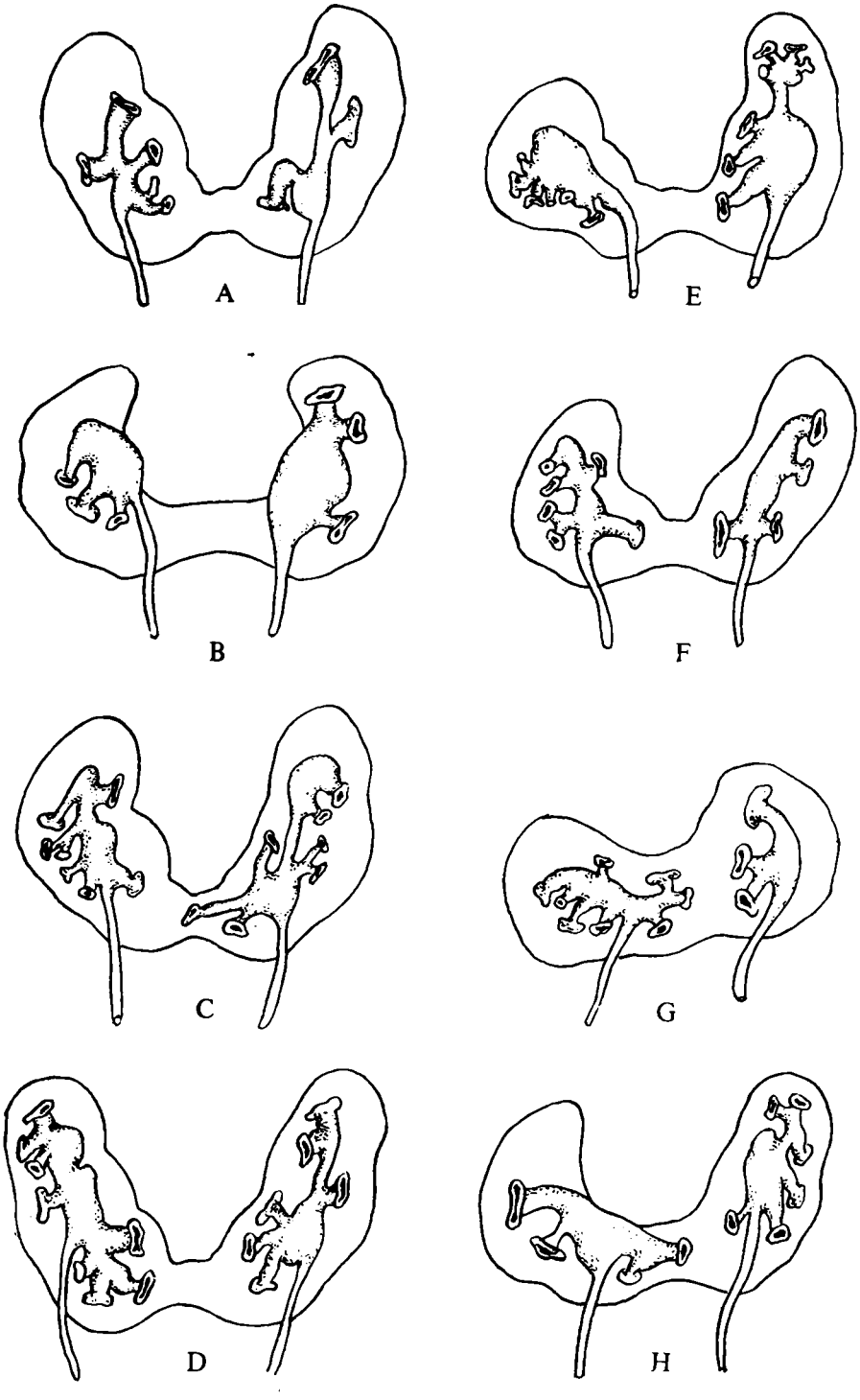


Fig. 32. Most common variants of fused kidney (drawn from pneumopyelograms).
A-H, horseshoe kidneys

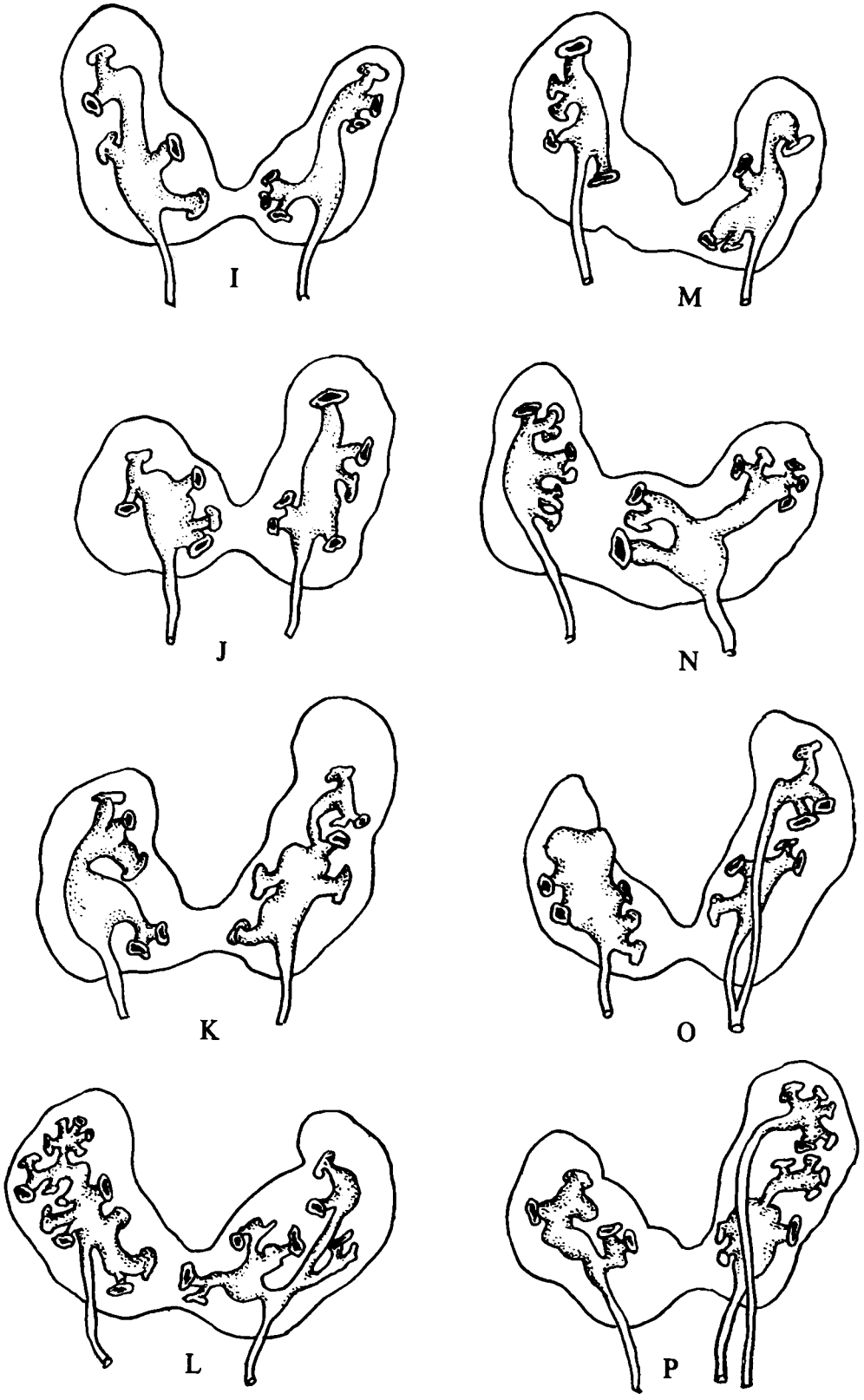


Fig. 33. Most common variants of fused kidney (drawn from pneumopyelograms).
I-*P*, horseshoe kidneys

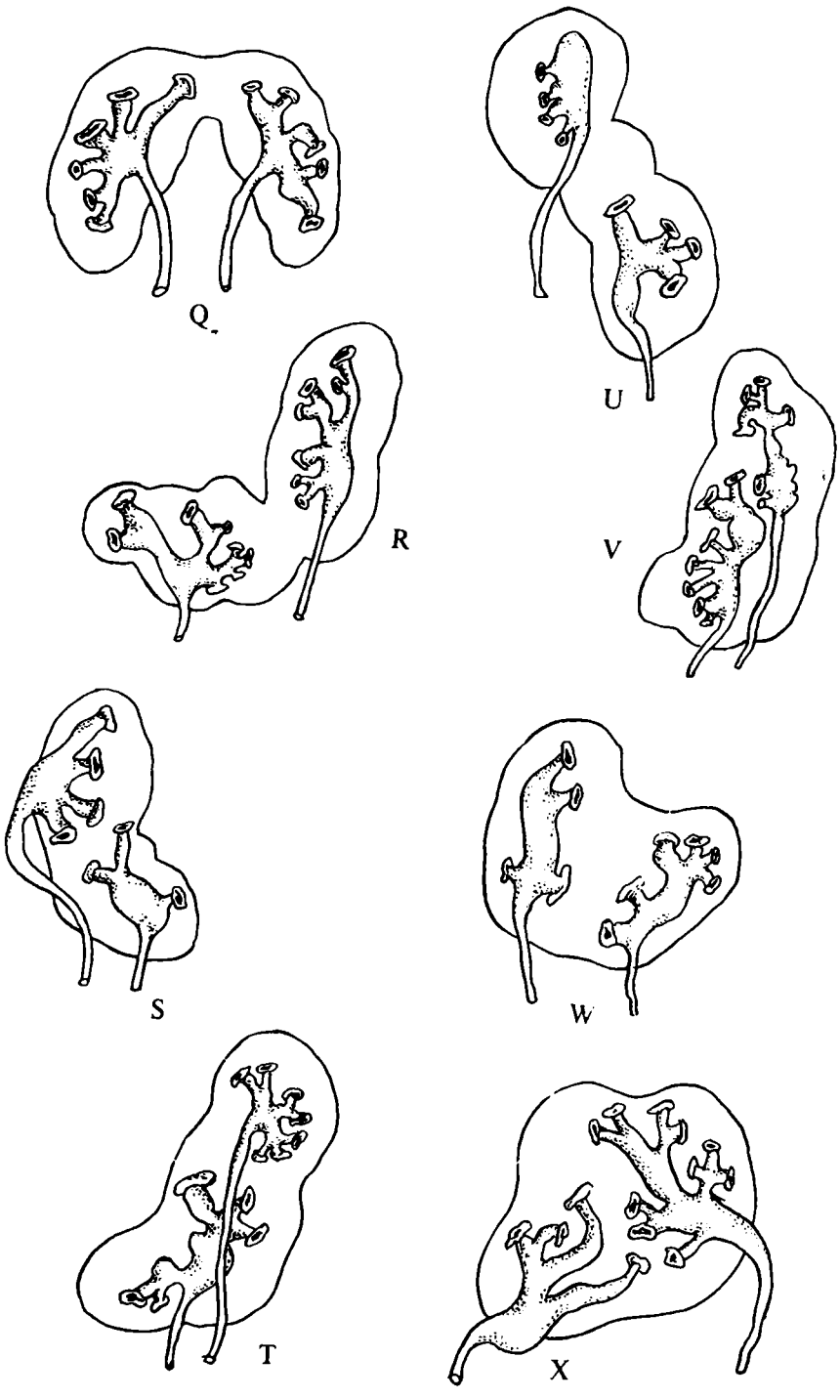


Fig. 34. Most common variants of fused kidney (drawn from pneumopyelograms).

Q, horseshoe kidney fused at the upper poles; *R-T*, angulated kidneys; *U*, sigmoid kidney; *V*, unilateral fused kidney; *W*, *X*, cake kidneys

isthmus can form from either both kidneys or from only one side. The line of fusion may be detected by the more or less conspicuous constriction on the anterior surface of the isthmus or a notch on its upper margin. Sometimes it coincides with the midline of the body; in other cases it is situated laterally, usually on the left. In a kidney with a large middle part the line of fusion cannot be determined.

The isthmus between the two kidneys may be of fibrous or parenchymal tissue. A fibrous isthmus is rare—we found it once only. Howard (1932) encountered it in three of 26 such kidneys and Zondek and Zondek (1964) in two out of 99.

In cases of a parenchymal isthmus the cortex of one kidney is continuous with the cortex of the other but the pyramids are separated (Vasilevsky, 1917). But if the isthmus contains only the cortical substance, then, according to Efremov (1913), the convoluted tubules are arranged in oblique rows and drain the urine into the straight tubules of the lateral parts.

In most cases the isthmus is 1 to 6 cm in length and width and 1 to 2.5 cm thick. The isthmus of a horseshoe kidney is commonly situated under the peritoneum above the aorta and vena cava on the level of the fourth to fifth lumbar vertebrae, but sometimes it is displaced upward, downward or laterally. Very rarely it can even be found to the back of or between the above mentioned vessels (Meek and Wadsworth, 1940; Aratsky, 1967).

The anterior surface of the isthmus is usually convex, the posterior surface concave. Due to this, and also to the numerous accessory vessels, the fused kidney is fastened tightly in its seat and cannot be drawn out into the wound during an operation. But even with a tightly fastened isthmus the lateral lobes of the kidney, its upper poles in particular, are slightly mobile.

Luchansky's (1957) research showed that the parenchymal tissue of the isthmus tends to undergo degeneration. Evidence of this is the poorly demarcated boundary between the cortical and medullary substances and the frequently encountered unclearly outlined oval glomeruli. In the middle part of the isthmus these glomeruli are often empty and hyaline casts are seen in them. This is not encountered in the lateral parts. The interstitial tissue is, however, well developed and there is more of it than usual. Thick bundles of smooth muscle fibres may often be randomly located here. All this is possibly a consequence of deficient nutrition and frequent inflammation.

The presence of the isthmus is evidently an explanation of the large weight and size of the fused kidneys. Two normal human kidneys weigh 240 g on the average, whereas the weight of fused kidneys described in the literature ranges between 300

and 460 g. Popov (1936) claims that fused kidneys are rarely smaller than normal kidneys.

The size of the isthmus affects the shape of a horseshoe kidney. The lateral lobes of such a kidney are nearer to the vertebrae than normally. The position of the longitudinal axis of each kidney varies greatly. Usually the axes pass downward from a lateral toward a medial point and intersect caudally, but when the isthmus is long they run parallel to each other. The asymmetrical position of the isthmus often causes the upper pole of the left half of the kidney to be located higher than that of the right half. In some cases the poles of such a kidney are on the same level, but in others the upper pole of the right kidney is situated higher than the upper pole of the left. This is probably associated with the size and position of the liver, which may also contribute to the large size of the left half of the kidney.

The shape of a fused kidney is also determined by the configuration of the fused parts, which are rarely classical in shape. They are usually elongated, spherical or elliptic; sometimes their shape is non-descript.

The anterior surface of the fused kidney is usually irregular, the posterior surface being smooth. Both lower poles of a horseshoe kidney with a long isthmus have vertical grooves and impressions resulting from the pressure of the large vessels.

The position of a fused kidney is linked with the extent and way of fusion. It is situated lower than a normal kidney, usually from the level of the first lumbar vertebra downward. In asymmetrical fusion, i.e. in unilaterally fused and sigmoid types, the organ may be located higher. The upper pole in this case will be found at the normal level. The lower kidney is usually displaced to the contralateral side; Foley (1940) encountered this in 92 per cent of unilateral fused kidneys. In our cases the right kidney was located on the contralateral side just as often as the right.

The **urinary tract** of fused kidneys is also unusual. The hilus may be poorly developed or not at all. Bogdansky's (1950) research showed that a hilus situated on the anterior surface of the organ appears as a shallow depression with a flat bottom, but one located on the inner border is a narrow and deep groove which comes close to a normal hilus in shape. These are the two extreme types, but intermediate shapes may be found in which it is easy to note the same regular features. In most cases the hili are located asymmetrically. They may be ventral on one side and be in the normal place on the other. The left hilus is usually nearer to the anterior surface of the kidney.

Depending on the position of the pelves, Fedorov (1923) distinguished three types of horseshoe kidneys. The first type, which

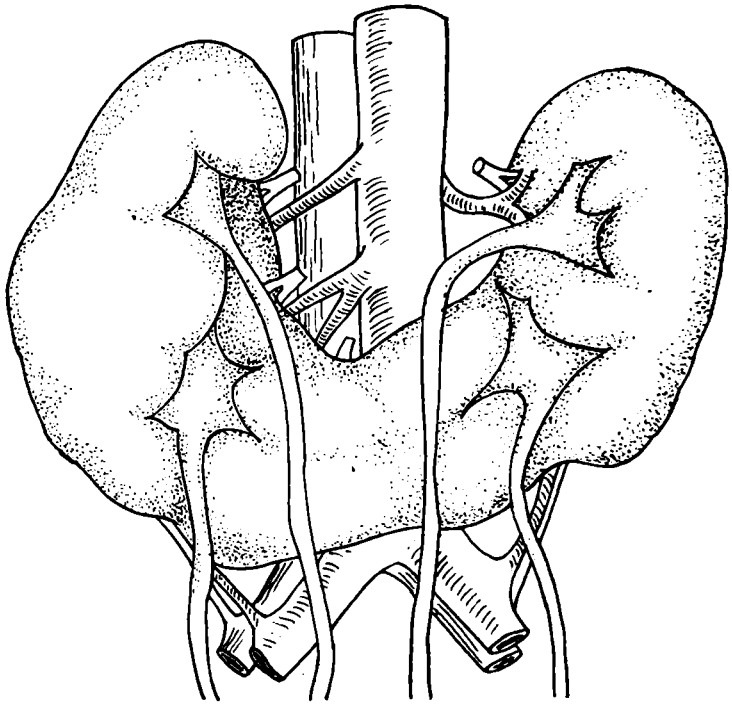


Fig. 35. Horseshoe kidney with complete bilateral duplication of the urinary tract (from postmortem examination)

most closely approximates a normal kidney, has extrarenal pelves located on the anterior surface. The second type has intrarenal pelves which are also more to the front. In the third type, one or both pelves are on the median or even on the posterior surface of the kidneys.

The pelves of a unilateral fused kidney are situated one below the other, whereas those of a sigmoid kidney face the opposite sides, the upper pelvis is usually directed medially and the lower pelvis laterally.

Fused kidneys have two separate pelves and two ureters. Duplication of the upper urinary tract on one or both sides in such cases, however, is not a rare occurrence. We encountered duplication in our practice in 18 cases (on the right in ten and on the left in eight). The duplication was total in three patients and partial in 15. In one of the patients the ureter was in a retrocaval position. Total bilateral duplication was encountered in only one of our cases (Fig. 35).

Accessory pelves are sometimes situated in the isthmus of a horseshoe kidney. In such cases when the isthmus is massive it seems as if there are three fused kidneys separated by grooves.

With such an anomaly the calyces are usually more numerous and unusual. They may be found inside the kidney, above, below,

medial to the pelvis or outside the kidney on its anterior surface. We encountered extrarenal localization of the calyces in seven clinical observations and in two postmortem examinations.

The calyces of a fused kidney sometimes cross the isthmus and terminate in the contralateral half of the kidney. For instance, in a case described by Potampa et al. (1949) both ureters were connected with the left half of the kidney where both pelves were located, but a calyx arose from one pelvis and formed an independent pelvis in the right half of the kidney. Cases described in the literature with calyces located in different pelves but connected with each other are evidently of secondary origin.

According to Bogdansky (1950), in 50 per cent of cases the ureters originate from the pelves at a higher level than normal and, most important, at a sharp angle. They are usually located on the anterior surface of the organ, but cases have been described when the ureters were found within or behind the isthmus (Kinard, 1923).

Shorter ureters of horseshoe kidneys often curve characteristically in the upper portion. In asymmetrical types of this anomaly one of the ureters transects the midline. The distal ends of the ureters of fused kidneys are usually found in their normal sites, although cases have been described of the right ureter draining into the left side of the urinary bladder and vice versa (Walters and Priestley, 1932). Displacement of accessory ureteral orifices into the urethra, sphincter vesicae or the genitals, and congenital stenosis, valves and diverticula of the orifices have been described (Krymov, 1925; Abeshouse, 1943; Kuss and Grillet, 1959). Wilcox (1921) had a case in which a fibrous cord found in place of the ureter at the wall of the bladder created the impression that it was the only ureter.

The **vascularization** of fused kidneys is marked by the diversity in the number and routes of the vessels. The different variants are shown in Fig. 36.

Papin and Eisendrath (1927) found that two thirds of 223 horseshoe kidneys had three to five arteries with some kidneys having up to ten arteries. In exceptionally rare cases a single artery branching to each half of the kidney may be encountered.

It is characteristic of fused kidneys that they are supplied with blood from several sources. Arteries often arise from the aorta as in normal cases and along the whole length of its abdominal part; the more dystopic is the kidney, the lower is the level of the origin of the arteries. According to Khromova (1947), the main left artery is usually shorter than the right. Besides, accessory vessels are often encountered arising from the aorta and its branches (inferior mesenteric, internal iliac, spermatic, suprarenal, and median sacral arteries). In most cases they ex-

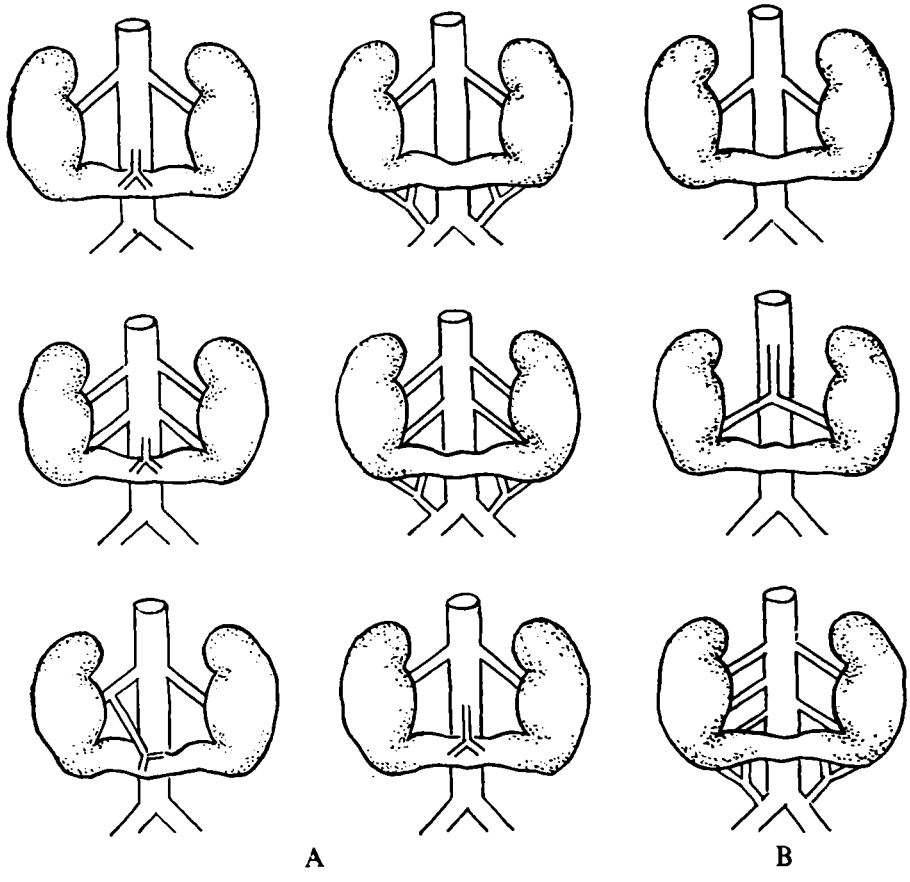


Fig. 36. Schematic representation of fused kidney vascularization. *A*, the most common variants (after E. Gimpelson, 1949); *B*, rarer variants

tend to the isthmus and lower poles of the kidney. Pepersack (1961) believes that in 25 to 65 per cent of cases the isthmus has its own artery. In 80 per cent of cases the number of vessels in each side of the kidney is not the same.

Most arteries approach the hilus of the kidneys from the back and above the pelvis. The main trunk stretches above the pelvis and the accessory branches behind it (Bogdansky, 1950). Sometimes the arteries are found above the pelvis. It should be borne in mind that in practice they may approach each lobe of a kidney that is separated by grooves or may drain directly into the kidney where there is no depression.

Efremov (1913) and Movshovich (1954) proved by angiography that the vessels of both halves of a horseshoe kidney communicate directly in the isthmus; Kazartsev (1967) pointed to the segmentary structure of this anomaly. Our observations also confirmed this. The borders of the vascular segments usually coincide with the grooves on the surface of such a kidney.

The veins in most cases correspond to the arteries in number and follow their course. Sometimes, however, their number may be less than that of arteries but still greater than that in a normal kidney. They drain into the vena cava inferior, sometimes into the common iliac, internal iliac, spermatic, phrenic or middle sacral vein. Veins of other organs may drain into the vein of a fused kidney. In a case observed by Bikmukhametova (1963), for instance, the suprarenal and inferior mesenteric veins and a branch of the splenic vein drained into the left vein of a horseshoe kidney. In such a case ligation of the renal vascular pedicle at a distance would have resulted in disturbance of the blood flow from the adrenal gland and intestine.

The veins entwine most of the renal pelvis. The thickest trunk usually stretches in front of it, covering the upper or middle third, while the accessory veins are behind. Thus, only the lower part of the pelvis is free of vessels (Bogdansky, 1950).

Judging from Vishnevsky's studies (1929), horseshoe kidneys are **innervated** in the same way as normal kidneys. The nerves arise from the trigeminal, superior mesenteric, renal or aortico-renal ganglia, between which there are many networks and which have small and large branches to the vagus nerve. This nerve plexus closely connects such a kidney with all the internal organs. According to Valker (1941), the nerve plexuses of a horseshoe kidney are very powerful, which is particularly evident after the examination of normal kidneys; the nerve network is very thick, clearly visible and there are some trunks which are abnormally thick.

Gutierrez (1931) pointed out that the **lymphatic system** of a fused kidney consists of a great number of lymph nodes joined to form lymphatic plexuses at the vessels. They are connected with the iliac and aortico-lumbar lymph nodes, thus forming the thoracic duct and lymphatic cisternae. It is possible that an anomalous kidney compresses this lymphatic system and impairs the normal circulation of lymph.

TOPOGRAPHICAL FEATURES

The fused organ is embedded in fatty tissue which is surrounded by renal fasciae. The isthmus of a horseshoe kidney does not have a fatty seat. The prerenal fascia and the peritoneum are in front of it.

With changes in the shape and position of an anomalous kidney the position of other anatomical structures interrelated to it also changes. This has been well studied in the case of horseshoe kidneys (Lisovskaya and Petrova, 1941; Khromov, 1947; Mardershtein, 1950).

A horseshoe kidney in a normal position hardly differs from

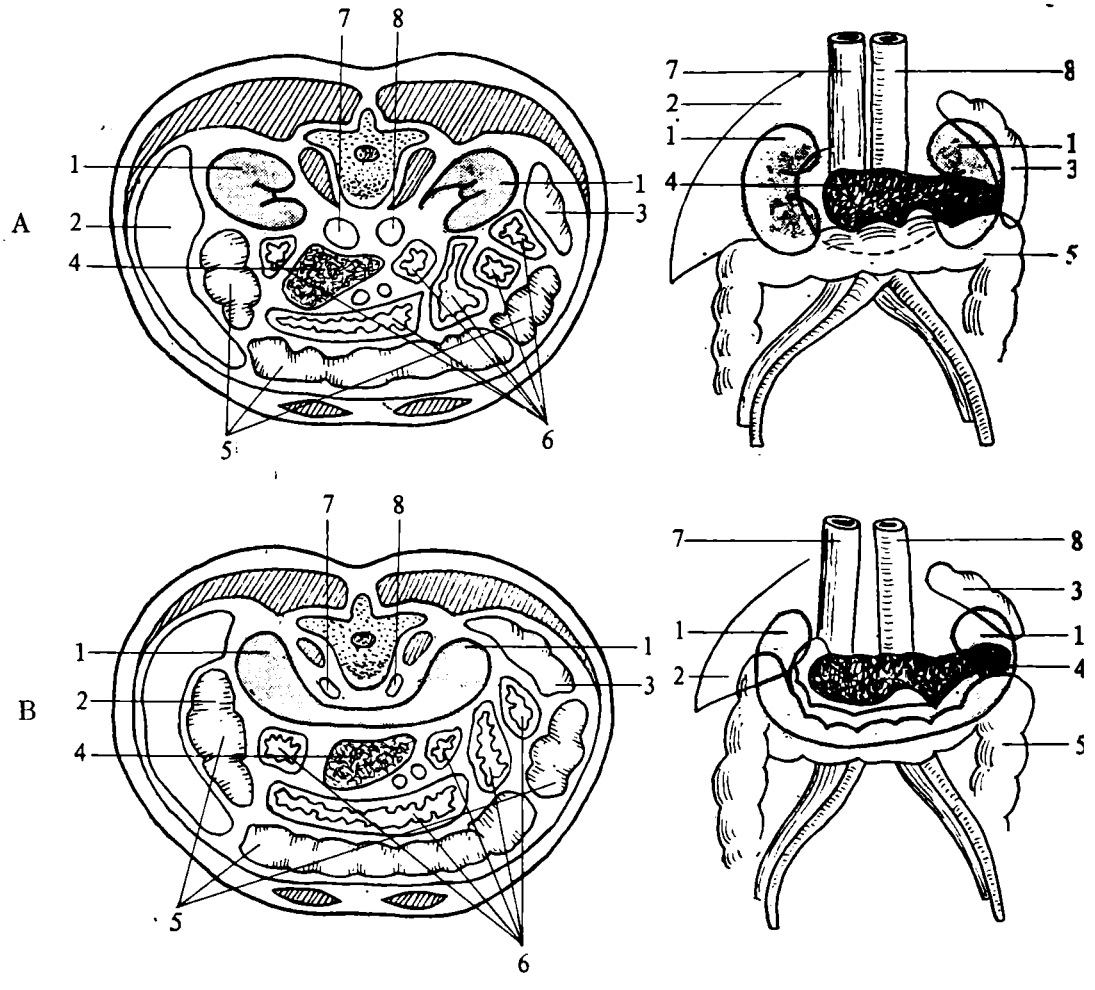


Fig. 37. Position of kidneys in relation to the abdominal organs (horizontal section at the level of the first lumbar vertebra).

A, normal kidney; *B*, horseshoe kidney. *1*, kidney; *2*, liver; *3*, spleen; *4*, pancreas; *5*, large intestine; *6*, small intestine; 7, vena cava inferior; *8*, aorta (after Khromov, 1947)

a normal kidney in its relation with other organs, but this is a rare variety. A horseshoe kidney is often encountered at the level of the second to fifth lumbar vertebrae. The position of the abdominal organs in such cases is shown in Fig. 37.

The position of the liver in these cases is not horizontal but almost vertical. Its left lobe occupies the seat of the right kidney, while the right lobe descends below the costal margin and its convex surface faces the front. The gall bladder is displaced laterally. The spleen is in the seat of the left kidney. The pancreas is often displaced forward and to the left. The distance between its head and the descending part of the duodenum is increased. The adrenals occupy their normal place in most cases,

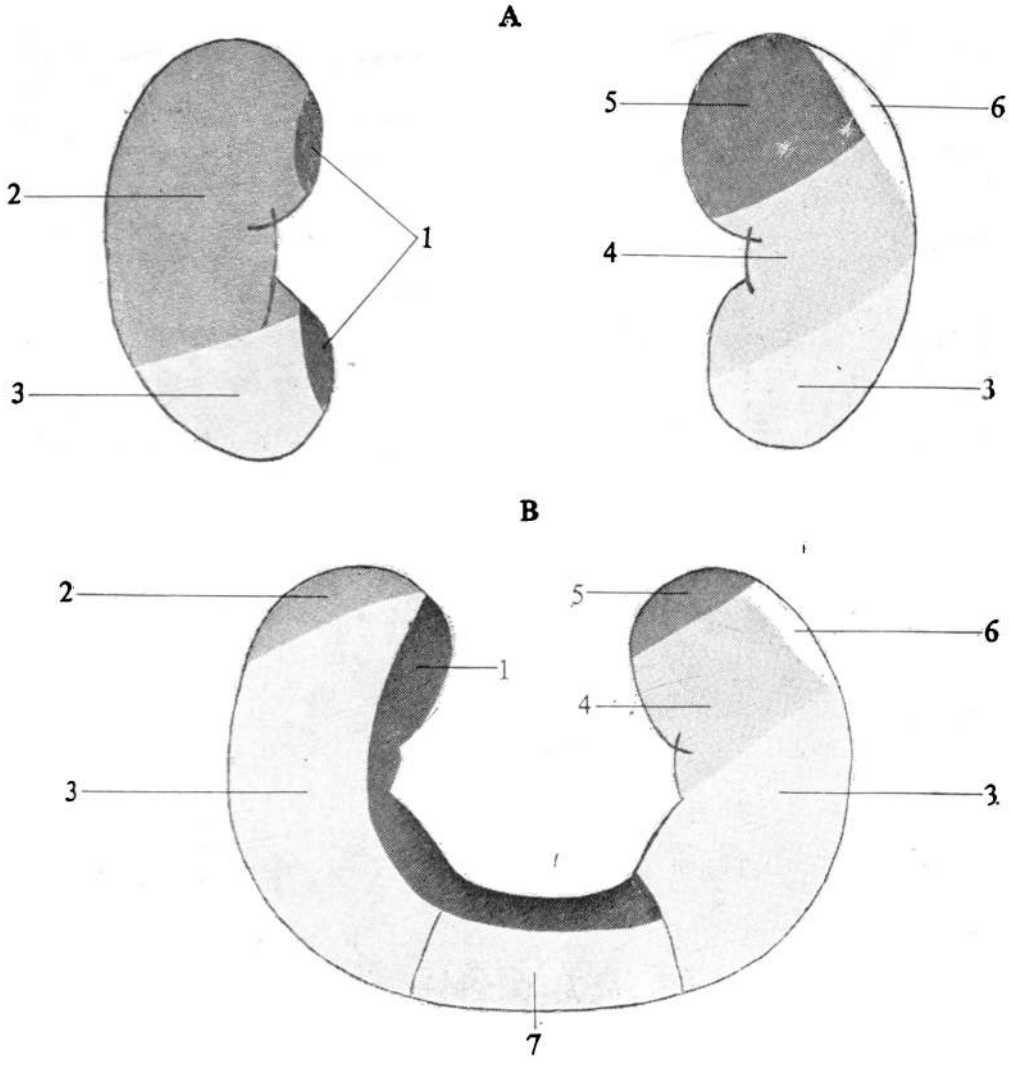


Fig. 38. Areas of contact of the anterior surfaces of horseshoe kidney with the adjacent organs (after Khromov, 1947)

A, normal kidney; *B*, horseshoe kidney. *1*, facies duodenalis; *2*, facies hepatica; *3*, facies colica; *4*, facies pancreatica; *5*, facies gastrica; *6*, facies lienalis; *7*, facies jejunalis

but sometimes may be found lower than normal, in which case they may be fused (Kelly and Burnam, 1914).

The hepatic flexure is displaced to the back, medially and upward. The transverse colon is far below its normal level and may be Y-shaped. That is why there is a kink in the region of the hepatic flexure. The descending colon is slightly displaced to the front. The duodenum deviates to the right and is pressed against the renal isthmus by the superior mesenteric artery. Its descending part is often displaced to the right, and the ascending part to the front and to the left, less frequently to the front and to the right. A strangulation groove is often seen on its horizontal

part from being compressed by the mesenteric vessels stretching over the renal isthmus. Up to this groove the duodenum may be dilated, which clearly indicates that movement of the contents is retarded here.

The aorta and vena cava in the region of the renal isthmus deviate laterally. Sometimes the aorta is wider above the isthmus than below it, in which case the left ventricle of the heart is hypertrophied. The superior mesenteric vessels arise from the aorta some distance above the renal isthmus and stretch in front of the duodenum. The inferior mesenteric artery originates directly above the isthmus.

The abnormal position of anomalous kidneys alters the areas of their contact with adjacent organs (Fig. 38). Organs corresponding to the anterior surface of the left kidney are: one third of the duodenum (which in most cases covers the upper half of the isthmus), the branches of the inferior mesenteric artery, and the pancreas which stretches along the superior margin of the kidney. The anterior surface of the right kidney corresponds with the inferior surface of the liver at the top (but it is not as close to it as a normal kidney), with part of the duodenum, and with the large intestine. The lumbar muscles and the diaphragm, the aorta and the vena cava correspond to the posterior surface of the lateral halves. A fused kidney located in the pelvis has areas of contact with the adjacent muscles, the iliac vessels, intestine, and the the uterus.

COMBINATION OF FUSED KIDNEY WITH DEVELOPMENTAL ANOMALIES OF THE KIDNEYS AND OTHER ORGANS

Fused kidneys often occur together with other developmental anomalies of the kidneys and urinary tract. As was pointed out above, the anomaly is found in combination with duplication of the upper urinary tract, hypoplasia of one half of the kidney (Judd et al., 1922), and multiple or solitary cysts in the kidney (Rathbun, 1924; Vorobtsov, 1956). Among 33 patients with a duplicated urinary bladder, three had a fused kidney (Burns et al., 1947). Zondek and Zondek (1964) found a horseshoe kidney to be combined with other anomalies of the urinary system in 15 out of 99 autopsies.

There are many reports in the literature on a fused kidney and concomitant genital anomalies: cryptorchidism (Meek and Wadsworth, 1940), absence or deficient development of the vagina and uterus (Byalyi, 1955), atrophy of the prostate and absence of the seminal vesicles (Popov, 1936). Reveno and Palubinskas (1966) found that six out of 40 patients with genital anomalies had a horseshoe kidney.

The incidence of a fused kidney and concomitant developmental anomalies of the kidneys and genitals is, apparently, in fact higher than is diagnosed. It is our experience that the various anomalies of other organs are diagnosed much less frequently among patients at the clinic than at autopsy. We recognized these conditions in 18 out of 206 patients, but found them in 5 out of 13 autopsies. This suggests that some of the anomalies of other organs remain unrecognized in the clinic.

Fused kidneys are sometimes combined with developmental anomalies of other organs which are so remote that a connection between the abnormalities cannot be established. Fused kidneys have been encountered in atresia of the rectum or the small intestine, fusion of both adrenals, absence of an adrenal gland, kyphoscoliosis, congenital vesicovaginal fistula (Gutierrez, 1934), diaphragmatic hernia (Roberts, 1931), vertebral defects like spina bifida or the presence of six lumbar vertebrae (Roberts, 1961), persisting thymus gland, lobulated spleen, Meckel's diverticulum (Ognev, 1929), and situs inversus viscerum (Mikhelson, 1959). Zondek and Zondek (1964) performed autopsies in 99 cases with horseshoe kidneys and found anomalies of the central nervous system in 19 of them, of the cardiovascular system in 18, of the respiratory system in 2, of the gastro-intestinal tract in 17, and of the skeleton in 16. The frequent combination of such anomalies can clearly be explained by the high incidence of developmental anomalies in general. This is confirmed by Humphry and Munn (1966), who found that the frequency of renal anomalies among 400 children with cardiovascular anomalies was approximately equal to that among the same number of children with a healthy heart.

STATISTICS

Fused kidneys are not as rare as was previously thought. Up to 1912, Botez was able to find only 320 cases in the literature, among which 270 were accounts of postmortem findings. In contrast, in the following years we found in the available literature reports on 1724 such anomalies, 549 of these reports dealing with autopsy cases. In the Soviet literature, 542 cases of fused kidney (253 from autopsies) have been described. Thus, the number of cases of fused kidneys described in the literature exceeds now 2000.

Most publications deal with individual cases, but special note should be made of the autopsy statistics reported by Sokolov (1928), Zamyatin (1931), and Zondek and Zondek (1964), accounting for 40 to 99 cases and the clinical statistics (Zeiss and Boeminhouse, 1934; Gibert, 1958; Glenn, 1959; Akhundov, 1968)

Table 1. Frequency of Fused Kidneys According to Autopsy Findings

Author	Number of autopsies	Number of fused kidneys	%
Gubarev (1892)	12 418	9	0.07
Efremov (1913)	3697	16	0.43
Arkhangelsky (1926)	3906	14	0.35
Sokolov (1928)	50 198	66	0.13
Chunovsky (1929)	2572	3	0.11
Zamyatin (1931)	13 547	40	0.29
Howard (1932)	14 146	22	0.15
Jeck (1932)	16 735	26	0.15
Shoemaker and Braasch (1939)	13 475	35	0.25
Abeshouse (1944)	1100	5	0.45
Nation (1945)	15 728	32	0.21
Bazhanov (1949)	1700	5	0.29
Shimkus (1969)	5596	12	0.21
Total:	154 818	285	0.18

embracing 36 to 51 cases. Culp and Winterringer (1955) published information on 106 surgical cases treated in their clinic over a period of 50 years. We possess data on 206 clinical cases and 12 autopsies.

The frequency of fused kidneys can be judged from the autopsy findings (Table 1).

It can be seen from the table that fused kidneys are found, on the average, in 0.18 per cent of autopsies (from 0.07 to 0.45 per cent).

In the clinic the anomaly is encountered more frequently.

Fused kidneys account, on the average, for 0.30 per cent (from 0.11 to 5.4 per cent) of all cases of diseases of the kidneys and upper urinary tract (Table 2).

Among the patients treated at our urological clinic, 1.3 per cent had fused kidneys; the anomaly was revealed in 2.6 per cent of patients during operation and in 0.7 per cent of those examined by excretory urography or retrograde pyelography.

There are different points of view on the sex predisposition of the anomaly. Some works point out the predominance of females among people with fused kidneys (Davidson, 1941; Vourinen, 1962); others note the opposite (Lowsley, 1952; Glenn, 1959).

Table 2. Frequency of Fused Kidneys in Patients with Diseases of the Kidneys and Upper Urinary Tract

Author	Number of patients	Number of fused kidneys	%
Bugbee and Losee (1919)	1000	4	0.40
Vorobtsov (1936)	1792	5	0.27
Abeshouse (1944)	3684	10	0.27
Smith and Orkin (1945)	18 460	24	0.13
Gimpelson (1949)	4549	35	0.76
Lowsley (1952)	13 080	46	0.35
Pekarskaya (1955)	925	6	0.64
Petkovic et al. (1961)	26 197	29	0.11
Kubarikov (1962)	240	13	5.40
Sokolyanskaya (1963)	400	12	3.00
Shimkus (1969)	4747	111	2.30
Total:	75 074	295	0.30

During autopsies Zondek and Zondek (1964) found that twice as many males as females had the anomaly, whereas according to Sokolov's (1928) statistics, fused kidneys were found in 0.13 per cent of autopsies on males and in 0.12 per cent of autopsies on females.

Our observations do not allow us to note any special predisposition to the anomaly of either males or females. We found fused kidneys in 1.33 per cent of our male patients and in 1.38 per cent of females.

Fused kidneys are predominantly revealed in patients in the 20 to 50 age group. Individuals of this age group constitute 57.6 per cent of all cases with the anomaly found on autopsy and 70.7 per cent of those recognized clinically by Sokolov (1928) and Culp and Winterringer (1955). In our clinical practice, 69.3 per cent of patients were of this age. The high frequency of diagnosed fused kidneys at this age is apparently linked with the heavy demands placed on the organism during intensive work as well as with the period of time needed for the defects of the anomaly to induce a disease. The number of individuals in whom the anomaly is revealed at an older age decreases gradually, and some apply for examination because of a kidney ailment at an old age, while in others a fused kidney is a chance finding at autopsy.

This anomaly is often revealed in very young children, evidently due to the fact that severe concomitant developmental

anomalies of other organs are diagnosed at an early age which make examination of the child necessary.

Fused kidneys are among the most commonly encountered kidney anomalies. They accounted for 206 (14.6 per cent) of 1405 various developmental abnormalities of kidneys encountered in our practice. In frequency they come second after duplicated kidneys.

Horseshoe kidneys fused at the lower poles predominate among autopsy findings and clinical cases (84.5 per cent). The remaining types are distributed according to frequency in the following order: angulated, cake, unilateral fused, horseshoe kidneys fused at the upper poles, and sigmoid kidneys.

Fused kidneys often cause various disorders and diseases. In our experience, 95.5 per cent of fused kidneys were the cause of disorders, while 4.5 per cent were asymptomatic and were detected only during examination.

Hydronephrosis, nephrolithiasis and inflammatory diseases predominate (about 90 per cent) in individuals with this anomaly. This is in agreement with data published by Culp and Winterringer in 1955 (87 per cent). The actual number of inflammatory diseases in this anomaly is apparently higher; only laboratory tests prove the presence of fused kidneys in some cases and not all patients undergo surgery.

A combination of these diseases in half of the kidney or bilateral affection of the whole fused kidney is often encountered. This special predisposition of the anomalous condition to hydronephrosis, nephrolithiasis and pyelonephritis, to the combination of these diseases in one kidney, and to bilateral diseases is apparently linked with the anatomical defects of the fused organ which are conducive to the stasis of urine in the upper urinary tract and disturbances of the circulation of lymph and blood.

Otehr diseases which are not associated with disturbances in the flow of urine are relatively rare in individuals with fused kidneys. This is also confirmed in the literature. For instance, Gimpelson (1949) encountered them no more frequently than in individuals with normal kidneys. Pepersack (1961) claims that the anomaly has a certain predisposition to the development of tumours and tuberculosis.

SYMPTOMATOLOGY, CLINICAL COURSE AND DIAGNOSIS OF FUSED KIDNEYS

Fused kidneys not complicated by diseases often cause various disorders which indispose the patients to some extent.

Pain is the most constant sign. It is sudden and is mainly experienced when the patient is in a vertical position, when he

turns the trunk sharply, during walking or physical exertion. The pain is relieved when the patient lies down and rests fully. A diet does not alleviate it.

The pain may be dull and continuous or acute. An attack of acute pain lasts several minutes, hours or days and is then followed by a clear period. Depending on the position of the anomalous organ, pain is located in different parts of the abdomen: the epigastric, paraumbilical, iliac or suprapubic region, on one or both sides of the hypochondrium or the lumbar region. Sometimes pain is felt in the whole abdomen, and is referred to the iliac region, the lumbar region, the lower limbs, the genitals, the urinary bladder and the rectum.

We sometimes observed in patients with a horseshoe kidney a more characteristic localization of pain. The following case is given in illustration.

S., a 56-year-old woman, was admitted to the clinic on June 6, 1962. She complained of dull pain in the region of the umbilicus experienced periodically over the last 10 years. An elastic, painless and hardly mobile tumour was palpated below the umbilicus. It was felt as an elevation lying across the spine and extending toward the hypochondria. The tumour transmitted the aortic pulse, and a systolic murmur was heard when a stethoscope was pressed against it. Rovsing's sign was positive. Blood test and urinalysis showed no abnormalities. Excretory urography demonstrated the shadow of the left pelvis on a level with the third to fourth lumbar vertebrae. Retrograde pyelography showed the right pelvis above the fourth lumbar vertebra. Diagnostic pneumoretroperitoneum produced the image of a horseshoe kidney, which verified the diagnosis finally. There were no signs of kidney disease. This was confirmed by a 6-year follow-up of the patient.

The umbilical localization of the pain in this case was not characteristic of a kidney lesion. The discovery of a characteristic tumour in this region fixed our suspicions on a horseshoe kidney. The combination of these symptoms is very important in the diagnosis of a fused kidney.

Pain in cases of fused kidneys is evidently caused by compression of the nerve plexuses by the malpositioned organ, although Gutierrez (1932) believes that the pain occurs as a result of an enlarged kidney that is excessively filled with blood.

Rovsing (1911) considered the development of pain in the abdomen on sudden bending of the trunk to be a characteristic symptom of a horseshoe kidney. This symptom, however, is very rare and may be found in some patients with other diseases who do not have this anomaly.

In some cases of very sharp pain Pasternatskii's symptom may be positive.

A **tumour** can quite frequently be palpated in the abdomen of patients with fused kidney. It is elastic to the touch, has a smooth or, less frequently, irregular surface, and is not very mo-

bile. It transmits the pulse of the aorta clearly. The tumour may be situated in different parts of the abdomen, but usually close to the vertebrae.

As is seen from the case described above, the tumour palpated in a patient with a horseshoe kidney has a characteristic shape. It lies across the spine and is felt as an elevation extending from the midline of the body upward and laterally. A fused kidney localized in the true pelvis may be revealed by vaginal or rectal examination.

If an elastic, painless, smooth and only slightly mobile tumour surrounded by large vessels is found on vaginal examination, the physician should bear in mind that this may be a fused kidney or a kidney displaced to the pelvis. This helps to avoid an unwarranted surgical intervention.

In asymmetrical types of fused kidney the tumour lies in the hypochondrium. It resembles a normal kidney in shape or else is more elongated.

Gregoir (cited in Pepersack, 1961) noted a characteristic symptom linked with a palpated horseshoe kidney: a coarse systolic murmur is heard through a stethoscope pressed against the tumour. This symptom, however, is not always present.

Disorders of the gastro-intestinal tract are rarely encountered. The manifestations are the same as those commonly observed in gastritis, perityphlitis, appendicitis or colitis. Our patients usually complained of constipation and less frequently of heartburn, hiccup, eructation, nausea, vomiting and meteorism. These symptoms are probably associated with pressure exerted by the anomalous organ on the nerve plexuses.

Neurological disorders are usually attendant to pain and gastrointestinal disturbances. They are rare and occur in the form of neurasthenia and neurovegetative dystonia. Patients complain of irritability, insomnia, malaise, indisposition and headache. In addition, some patients complain that they have a sensation of strong pulsation in the abdomen. Martynov (1910) claimed that this symptom is very distressing to the patient. He explained it by the swelling of the renal isthmus as blood fills the viscera.

Some patients with a horseshoe kidney experience paraesthesia in the abdomen and lower limbs. Gimpelson (1949) considered this symptom most characteristic of fused kidneys.

Disorders of the cardiovascular system are associated, according to many researchers (Pepersack, 1961), with pressure exerted by the isthmus of a horseshoe kidney on the aorta (coarctation of the aorta) and vena cava (Plate VIII), and are therefore typical of this anatomical type of fused kidneys. Increased venous pressure in the vessels of the pelvis and lower limbs, encountered

in cases of horseshoe kidneys, is conducive to the development of local extravasations and haemorrhages, cyanosis, oedema, phlebitis of the lower limbs, ascites, and, finally, thrombosis of the iliac vein and vena cava. In some cases compensatory hypertrophy and dilatation of the left cardiac ventricle, aortitis, dilatation or aneurysm of the aorta, and thrombosis of the aorta or its branches are explained by the permanent compression of the aorta by the horseshoe kidney.

We believe that the role of the horseshoe kidney in the development of severe changes in the cardiovascular system because of compression of the aorta or vena cava is overestimated. In our practice, compensatory left ventricular hypertrophy was noted in only 10.7 per cent of cases, and only one patient had increased arterial pressure. The associated occurrence of aneurysm of the abdominal aorta and a horseshoe kidney is rare (Love and Ross, 1968), and only a single case of thrombosis of the aorta and iliac arteries below the level of the isthmus of an anomalous kidney has been described (Hubbell, 1958). Comparison of the number of such complications with the total number of these diseases makes it apparent that the role of chance in their appearing together cannot be excluded.

Total haematuria may sometimes occur in individuals with a fused kidney but without kidney disease. We have had one such case.

S., a 41-year-old man, was admitted to the clinic on January 2, 1969 complaining of blood in the urine. He had been sick for three years, during which period sanguineous urine appeared three times without any apparent cause. Palpation revealed an elastic, painless and poorly mobile tumour in the region of the umbilicus.

Urinalysis: reaction acid, specific gravity 1020, protein 0.66 pro mille, leucocytes five or six in field of vision, erythrocytes covering the entire field of vision. Other blood tests and examinations of the urine showed no abnormalities. Careful urological examination did not reveal any disease. Pneumogram demonstrated the shadow of a horseshoe kidney.

In this case total haematuria was evidently associated with impaired flow of urine in the pelvis of the anomalous kidney and with compression of the vessels. Damage inflicted to the anomalous organ, which is less protected from physical factors, probably plays a certain role in the development of haematuria.

Chauvin and Chauvin (1954) and Pisano (1961) also observed total haematuria in individuals with a horseshoe kidney not complicated by disease.

Clinical Course

According to the clinical course, all patients with a fused kidney not complicated by diseases may be distributed into

two clinical groups: (1) without diseases or disorders and (2) without diseases but with disorders.

Individuals with asymptomatic fused kidneys are rarely encountered in clinical practice. Among our patients with fused kidneys only 19.3 per cent had no symptoms. The renal anomaly in these cases was an accidental finding during prophylactic examination or during X-ray examination for diseases of other organs.

Regular comprehensive follow-up of these patients over periods of one to 13 years showed that even later they had no complaints of any disorders in the kidneys, while the morphological and functional indices of the organs remained good. Nevertheless, even with such a relatively asymptomatic course a fused kidney causes some discomfort. The presence of a kidney in the lower abdomen called for X-ray examination and presented difficulties in its recognition and differential diagnosis.

In the other patients of this group we did find symptoms of a fused kidney. In some of them the first signs of the anomaly developed in childhood or adolescence, but in many they became evident much later, in middle or even old age.

The different periods at which the symptoms develop are possibly associated with the time necessary for the defects of the anomalous organs to be disclosed, as well as with loss of flesh, weakening of the organism as the result of systemic diseases, hard physical work, i.e. with factors which determine a change in the position of the kidney on the nerve plexuses.

Disorders in a case of fused kidneys usually trouble the patient for many years. They rarely occur suddenly, as was the case described below.

G., a 42-year-old man, was admitted to the surgical department of the clinic on January 3, 1959 as an emergency. He complained of attacks of acute abdominal pain, nausea, vomiting, and absence of stool and gases. Ileus was suspected and diagnostic laparotomy performed which established that the cause of the disease was an angulated kidney. After the operation the patient was transferred to the urological department. An elastic, painless, and hardly mobile tumour was palpated on the left and below the level of the umbilicus. Blood and urine tests showed no abnormalities. X-ray examination enabled the diagnosis of angulated kidney to be made.

When the disease is chronic, the first symptoms are followed by a clear period which is again replaced by a period of pain. In some cases the attacks of pain occur at small intervals, in others there may be clear periods of months or even years. Sometimes the attacks occur at shorter and shorter intervals and the intensity of the symptoms grows. The following case is an illustration of the chronic course of the anomaly.

Z., a 51-year-old woman, was admitted to the clinic on April 2, 1956. She complained of dull pain in the right half of the abdomen and right lumbar region. She had been ill for over 30 years, experiencing the pain at intervals of five to eight years. During the progress of the disease she underwent two operations, appendectomy and diagnostic laparotomy, which did not bring any relief. Postoperative scars are seen in the right hypochondrium and right iliac region. Blood and urine tests showed normal values. A horseshoe kidney was diagnosed on X-ray examination. No renal diseases were revealed. In a follow-up period of 13 years the patient experienced pain twice, each time for a duration of one month, but her condition in the periods between the attacks was good and no functional or morphological abnormalities were found in the kidney.

This case also illustrates how the long duration of the disease can lead to unwarranted surgical intervention.

Fused kidneys not infrequently trouble the carrier for many years. Most of our patients had noted disorders for five to 20 years or more.

Disorders due to fused kidneys are sometimes exacerbated during pregnancy. The anomaly, in turn, may aggravate the course of pregnancy and impair delivery (Anderson et al., 1951).

According to Bell (1946), pregnancy followed a normal course in only 62 per cent of 153 women with fused kidneys.

The course of the anomaly may be complicated by diseases of the adjacent organs and, vice versa, it may create difficulties in their treatment. The great number of arteries and the position of the isthmus of a horseshoe kidney cause additional difficulties and increase the risk of surgery for aneurysm of the abdominal aorta. In such cases the kidney has to be freed from the accessory vessels and its isthmus shifted from its place or transected (Falor and Rufflo, 1964).

In general surgical and gynaecological practice the clinical manifestations of a fused kidney are often the cause of diagnostic errors and unjustified operations which sometimes have a tragic outcome. Reports have appeared at times in the literature of total removal of a fused kidney (Buss, 1899; Strube, 1905; Idel, 1927; Anderson and Harrison, 1965).

Gutierrez (1932) claims that sooner or later a fused kidney causes greater or lesser manifestations of active or passive disorders revealed or not revealed clinically. That is why, from the anatomical and clinical standpoint, and, as it will be seen below, the surgical standpoint as well, the name 'horseshoe kidney disease' for this condition is well-grounded.

Diagnosis

It is difficult to make a diagnosis of fused kidneys by routine clinical methods of examination, because the clinical picture is not characteristic and is poor in symptoms. The medical history

helps if the patient knows that he has an anomalous kidney.

The diagnostic difficulties usually occur in the first stage, the clinical examination yielding little to suggest the presence of a fused kidney. Complaints of characteristic pain at the umbilicus and an elastic, painless, and poorly-mobile tumour palpated at the vertebrae prove valuable in this respect. In 58 per cent of our cases this was enough to bring the anomaly to mind. The other symptoms are of slight diagnostic value. Laboratory and instrumental methods of examination do not demonstrate any abnormalities in this group of patients.

X-ray methods of examination play the principal role in the diagnosis of fused kidneys.

A **scout urogram** often provides an image of the fused organ. We encountered this symptom in one third of our cases.

The shadows of the horseshoe kidneys on the radiographs were large and were located at a low level and close to the vertebrae. Their longitudinal axes ran obliquely so that the lower poles met at the vertebrae. The shadows of cake and angulated kidneys were also large and were seen close to the vertebrae. In some cases the lower poles of such kidneys could not be seen clearly. Sometimes these symptoms were demonstrated more clearly on one side and less clearly on the other.

In his cases of horseshoe kidneys Frangois (1949-1950) saw a large spherical shadow on the X-rays with an indentation in the middle; it appeared as if the shadow 'saddled the spine'. The same sign, however, is encountered in cases of a pancreatic cyst or aneurysm of the abdominal aorta.

The suggestion made by Lhez and Fregevu (1960) deserves attention: in studying a scout X-ray one should be guided by the length of the longitudinal axis and the position of the kidney shadows in relation to the midline of the body. The longitudinal axis of a normal kidney is 70 to 80 mm long and its middle point is at a distance of 75 mm from the midline. In 50 per cent of horseshoe kidneys the axis reaches 200 mm or more in length, while the distance between it and the midline is often shortened.

The characteristic image of a horseshoe kidney on the X-rays allowed us not merely to suspect this anomaly in nine cases, but to make the diagnosis with assurance, although there was no evidence of kidney anomaly in the medical history and the kidney was not palpated. In most cases, however, these findings are not sufficient for making a confident diagnosis of a fused kidney, including confirmation or exclusion of disease, and revealing the functional and morphological features of the fused organ. That is why excretory urography has to be carried out in all cases.

Attention should be drawn to still another sign which enables

one to suspect a kidney anomaly. With a fused kidney a scout X-ray sometimes shows the liver in an unusual position, at a low level and vertical. It evidently occupies the seat of the kidney.

Excretory urography often yields sufficient information for making a proper diagnosis, when the kidney is functioning well. Valuable data are obtained concerning function, while the abnormal shape, size and position of the pelves and calyces suggest that the kidneys may be fused to form a single organ.

The following characteristic signs of the anomaly may be disclosed by means of excretory urography: duplicated image of the pelvis, arrangement of the calyces on both sides of the longitudinal axis of the pelvis and ureter, inclusion of the inferior calyces in the 'lumbar rectangle' (they sometimes resemble a pyeloureteral orifice), abnormal angle between the longitudinal axes of both pelves, reduced angle of the 'lumbar triangle', displacement of both pelves to one side of the spine, close arrangement of the pelves and, in particular, the calyces, with the ureters entering the urinary bladder at the normal site, arrangement of the ureters in the form of 'flower vase contours', and in some cases dilatation of the prepelvic portion of the ureter and forward curvature of the upper third of the ureter (Fig. 39).

The low position of the pelves of a fused kidney and their close proximity to each other and to the midline of the body are demonstrated particularly frequently by excretory urography. This is especially noticeable in the asymmetrical types of fused kidneys. In a horseshoe kidney with a fibrous isthmus, however, the pelves may be located far from each other, as in sigmoid kidneys, which is why the diagnosis of fused kidney cannot be rejected merely on the grounds of this sign.

Other signs of fused kidneys in excretory urography are found much less frequently, and their diagnostic value is therefore less. In this group we did not encounter the 'double image of the pelvis' described by Surraco (1927). This sign consists in the anomalous pelvis producing two shadows, one in front of the other, which determines the density of the combined shadow.

Macquet and Weneau (1952) noticed that the calyces of a horseshoe kidney are frequently included in the 'lumbar triangle' formed by lines drawn between the ends of the transverse processes of the first and fifth lumbar vertebrae. This occurs because the calyces of a fused kidney lie medially of the pelves. The importance of this symptom was emphasized by our cases and by Cieslinski's data (1960). However, in asymmetrical variants of a horseshoe kidney and in normally positioned pelves it may be negative on one or both sides.

Sometimes the inferior calyces of a fused kidney are found on both sides of the ureter and give a false impression of being

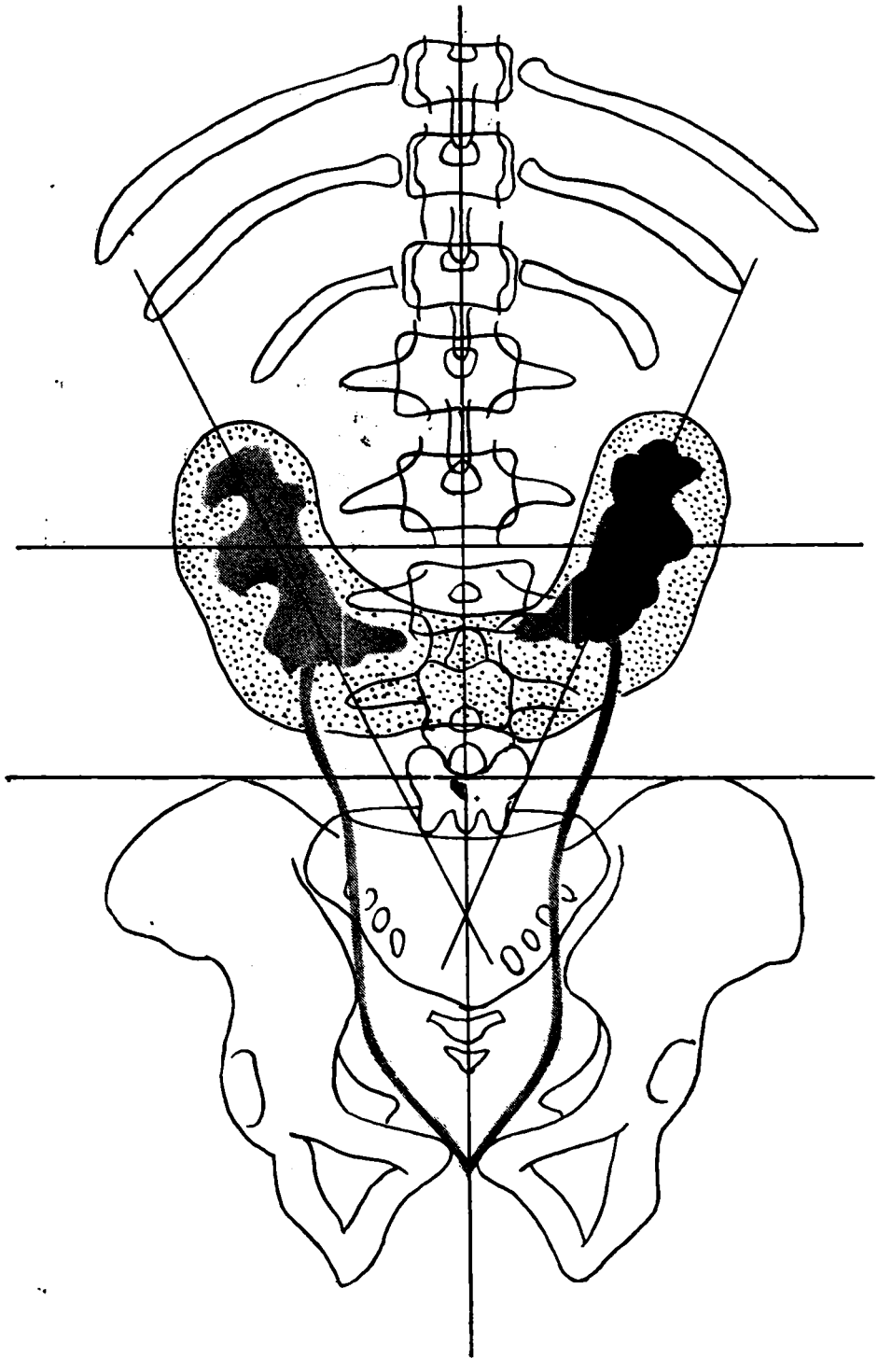


Fig. 39. X-ray signs of a horseshoe kidney in excretory urography or retrograde pyelography (discussed in the text)

the prepelvic part of the ureter. This part is dilated in some cases and resembles 'the neck of a bottle', as was noted for the first time by Gutierrez (1932).

The longitudinal axes of both pelves of horseshoe kidneys usually intersect caudally, forming an angle open upward. In cases of a long isthmus, the axis on one side or both axes may run parallel to the spine or the axes may be parallel to each other, no angle, therefore, being formed. The pelvic axes in a cake kidney are often parallel to each other; in an angulated kidney the longitudinal axis of one pelvis runs parallel to the spine, while that of the other pelvis is perpendicular to the first.

Gutierrez (1932) recommends determining the 'lumbar triangle', whose main angle in cases of a horseshoe kidney is smaller than that of normal kidneys. It is determined as follows. From a point in the middle of the fifth lumbar vertebra and on a line connecting the iliac crests, lines are drawn on both sides to the median surfaces of the inferior calyces. The sides of a triangle are thus formed, the base of which is a line drawn between the second and third lumbar vertebrae parallel to the line connecting the iliac crests. According to Gutierrez, the main angle of this triangle measures on the average 90 degrees (from 64 to 103 degrees) in individuals with normal kidneys, and 20 degrees (7 to 36 degrees) in cases with a horseshoe kidney. De Backer (1961) found that in children with a horseshoe kidney it was on the average 43 degrees (25 to 60 degrees). In our experience the main angle of the 'lumbar triangle' could be determined during excretory urography in only one third of cases. It was 50 degrees on the average (from 29 to 70 degrees).

The shorter ureter of a horseshoe kidney often has a characteristic position. On a posteroanterior radiograph both ureters stretch at first laterally, then curve medially, and finally again laterally and medially, thus resembling the outlines of a 'flower vase'. During excretory urography, however, the ureters are rarely seen full length.

In lateral-view radiographs the upper third of the ureters of a horseshoe kidney usually deviates forward, forming an arch over the isthmus (Renon, 1949). Because of the unclear image produced in excretory urography this symptom is rarely demonstrated.

With an adequately functioning kidney one would expect these symptoms to be demonstrated bilaterally, but in most cases they are in fact seen only on one side, which makes their diagnostic value still less. The reasons for this are the insufficiently clear image of the upper urinary tract produced on excretory urography and the frequent asymmetrical position of the anomalous organ. In a horseshoe kidney the middle of the isthmus usually lies above

the aorta and the whole kidney is consequently displaced to the left. The right pelvis in such cases is close to the vertebrae, sometimes above them. The shadow of the pelvis on the radiographs may thus be hidden in the shadow of the pelvic bones.

The diagnosis of a fused kidney is made most convincingly by excretory urography when both pelves and the image of the fused organ are shown on the radiographs (Fig. 40*A*).

Retrograde pyelography produces a clearer image of the upper urinary tract and therefore demonstrates the X-ray symptoms of fused kidneys more frequently (Fig. 40*B*).

The most common signs of fused kidneys revealed by this method of examination are: low position of the pelves, which are also seen to be close to each other; medial displacement of the calyces and their inclusion in the 'lumbar rectangle', with the inferior calyces closer to the spine than are the superior calyces; the main angle of the 'lumbar triangle' is smaller than 64 degrees; a short ureter, which often forms the outline of a 'flower vase'. It should be noted that the main angle of the 'lumbar triangle' could be determined in only 12 patients with horseshoe kidneys when the pelves were located above the level of the fifth lumbar vertebra. It measured 42 degrees on the average (from 24 to 70 degrees) and was more than 64 degrees only in two cases with a long renal isthmus. In most cases the pelves of the fused kidneys were more than 80 mm in length and the longitudinal axes of horseshoe kidneys fused at the lower poles formed an angle open upward. The other symptoms were rarely found.

On radiographs taken at an angle of 45 degrees the prepelvic part of the ureter was often displaced to the front because the ureter here stretched above the isthmus or the lower pole of the kidney. These radiographs show a better image of the calyces, which on a posteroanterior-view radiograph are hidden in the shadow of the pelvis.

The findings of excretory urography and retrograde pyelography, however, are not sufficient for diagnosing a horseshoe kidney when the calyces are directed laterally, even if only on one side. In such cases more precise information is gained by other methods of examination.

Because of the excellent contrast images, retrograde pyelography yields more precise information of the structure of the upper urinary tract of a fused kidney than does excretory urography. Even if the kidney of patients in this group functions adequately, excretory urography in many cases cannot demonstrate signs of the anomaly on both sides. This is particularly obvious on comparing radiographs made by these two methods of a single patient.

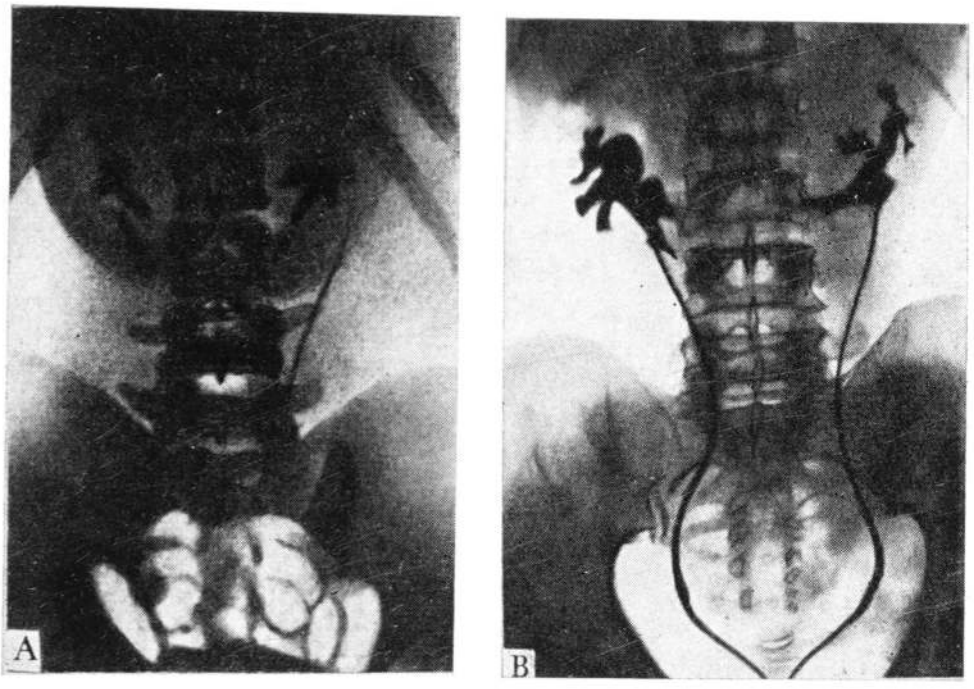


Fig. 40. Horseshoe kidneys.
A, excretory urography; *B*, retrograde pyelography

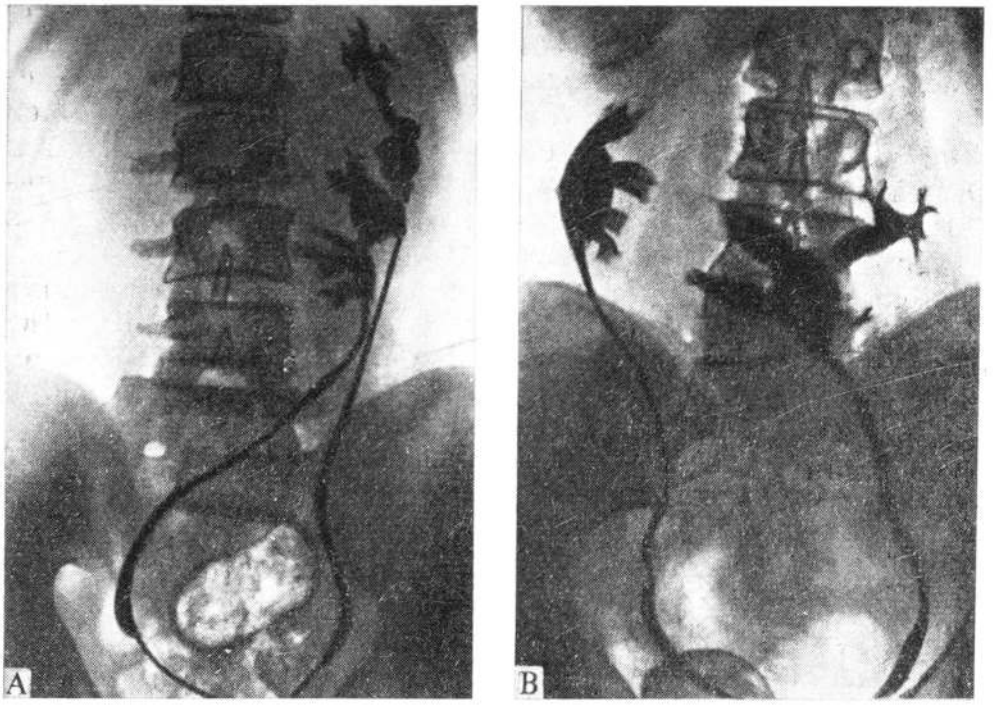


Fig. 41. Retrograde pyelography.
A, angulated kidney; *B*, unilateral fused kidney

Retrograde pyelography should be used in the diagnosis of fused kidneys when the results of excretory urography are not convincing, before an operation to reveal the details of the structure of the urinary tract, and also when it is necessary to exclude without doubt the presence of disease in the anomalous kidney. It should be borne in mind, however, that the signs revealed by retrograde pyelography are only indirect evidence of a fused kidney, even then when they are bilateral (Figs. 41*A*, *B*). Hence, bilateral examination is necessary; this is a shortcoming of the method because of the hazard of bilateral catheterization of the pelves. Besides, auxiliary methods of examination have to be used in doubtful cases.

Pneumoretroperitoneum allows the diagnosis of the anomaly to be made not from indirect signs, as is the case in the methods discussed above, but from the direct image of the fused organs on the radiographs. This makes the diagnosis more authentic (Fig. 42*A*). There are only a few reports in the literature on the use of this method in the diagnosis of fused kidneys, and they mostly deal with occasional successful cases (Hickel, 1959; Gluz, 1964).

The method provides a visual image of the anomalous kidney, its position in relation to the spine, and its size. It also demonstrates the type and extent of the fusion. On posteroanterior pneumograms horseshoe kidneys are seen as shadows lying obliquely in relation to the midline, with the lower poles hidden in the shadow of the vertebrae. The shadow of each half of the fused kidney on the pneumogram is on the average 12.5 to 13.5 cm long in men and 9.5 to 11.5 cm long in women, which is more than the average length of the shadow of a normal kidney. Diagnostic pneumoretroperitoneum makes it possible to examine the outlines of a sigmoid kidney (Fig. 42*B*).

Radiographs taken from the posteroanterior view do not give a clear image of the isthmus, whose shadow is concealed by the shadow of the vertebrae. But a good image can be obtained on radiographs made at an angle of 45 degrees (Fig. 43), in which case the area of the fusion is beyond the vertebral shadow.

The combination of the different methods (diagnostic pneumoretroperitoneum with excretory urography, retrograde pyelography or tomography) yields additional information (Fig. 44).

Pneumoretroperitoneum also helps in the proper diagnosis of a fused kidney when the anomaly is not immediately obvious because the pelves are far away from each other. It likewise makes it possible to differentiate the types of fused kidney. For instance, in one of our cases retrograde pyelography suggested an angulated kidney, whereas the pneumogram demon-

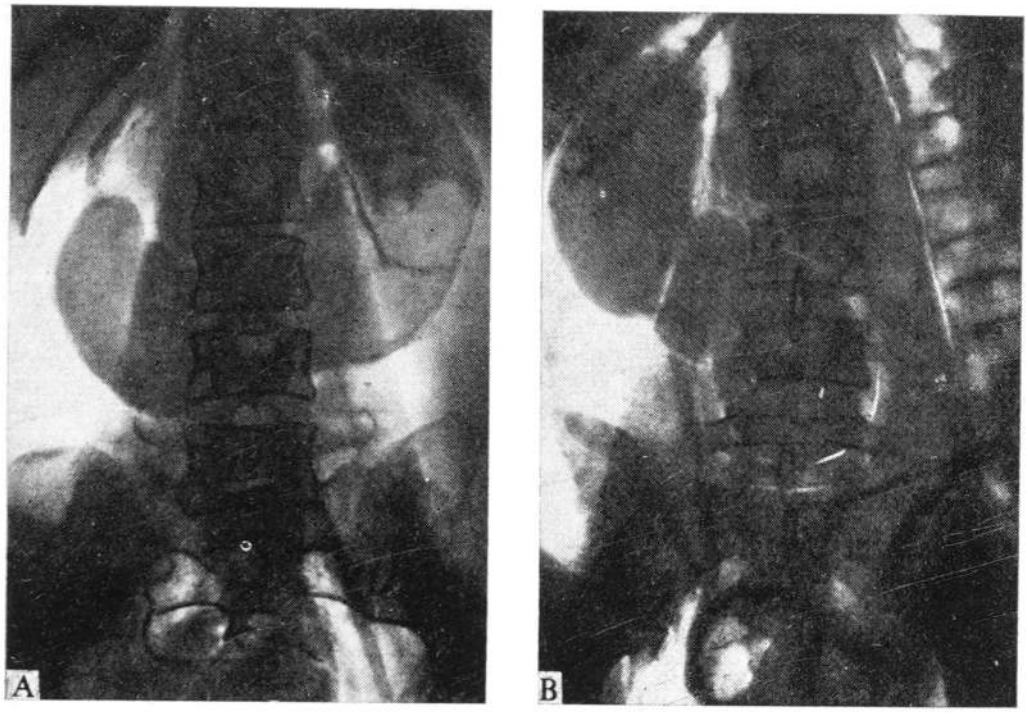


Fig. 42. Pneumoretroperitoneum.
A, horseshoe kidney; *B*, sigmoid kidney

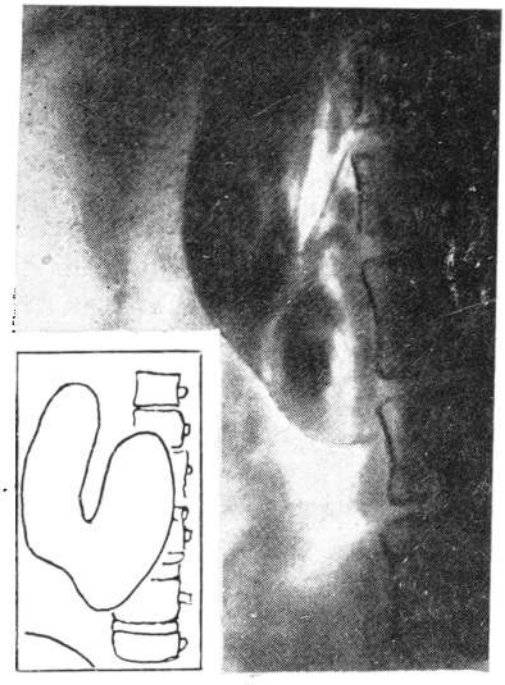


Fig. 43. Pneumoretroperitoneum. The radiograph is taken at an angle of 45 degrees; horseshoe kidney

Fig. 44. Pneumoretroperitoneum and excretory urography. Horseshoe kidney

strated a horseshoe kidney with a thick isthmus. In another case the findings of retrograde pyelography gave grounds for suspecting a unilateral fused kidney, but pneumograms made at an angle of 45 degrees allowed the proper diagnosis of an angulated kidney.

In only one of 13 patients in this group did diagnostic pneumoretroperitoneum fail to show an image of a fused kidney. This was a woman who had earlier undergone X-ray therapy for a tumour of the genitals. The gas did not penetrate the sclerotic perirenal tissues, as a result of which perinephritis was suspected as the cause of the pain. The findings of the pneumoretroperitoneum were also of diagnostic value in this case.

For the proper interpretation of the radiographs it is necessary to take into account the amount of air introduced into the retroperitoneum space and the degree to which it spreads in the space and around the kidney. It is our experience that to produce a clear image of the fused organ on the pneumogram 18 to 20 cm^3/kg of oxygen should be introduced.

We believe this method indicated in all doubtful cases, when it is difficult to prove or exclude fusion of the kidneys or when the type of the anomaly has to be specified.

In our experience, *tomography* as an independent method of examination for fused kidneys does not provide convincing data in cases in which scout radiographs have also failed to show an image of an anomalous organ. It has proved valuable as an auxiliary method combined with diagnostic pneumoretroperitoneum; it demonstrates those parts of the kidney which are unclear on pneumograms because of the superimposed additional shadows of the bones of the pelvis or spine. This is illustrated by one of our cases in which the results of retrograde pyelography suggested an angulated kidney, but pneumoretroperitoneum and tomography showed a horseshoe kidney with a thick and wide isthmus. Diagnostic pneumoretroperitoneum with tomography also help showing up cake kidneys on radiographs.

The height of the main tomographic section in our cases, with the patient in a supine position, was 9 to 14 cm.

Renal angiography may provide valuable information on the character of the blood supply to the fused kidney, and on its function, shape, and position. Lopatkin's (1961) research showed that the method is very important in the diagnosis of this anomaly and in its differentiation from tumours of the kidneys and other organs. Preoperative renal angiography gives an idea of the difficulties which may be encountered during an operation and the manipulations which will have to be undertaken, and suggests which arteries may be left intact and which may be ligated. Knowing the vascularization of the isthmus and the dis-

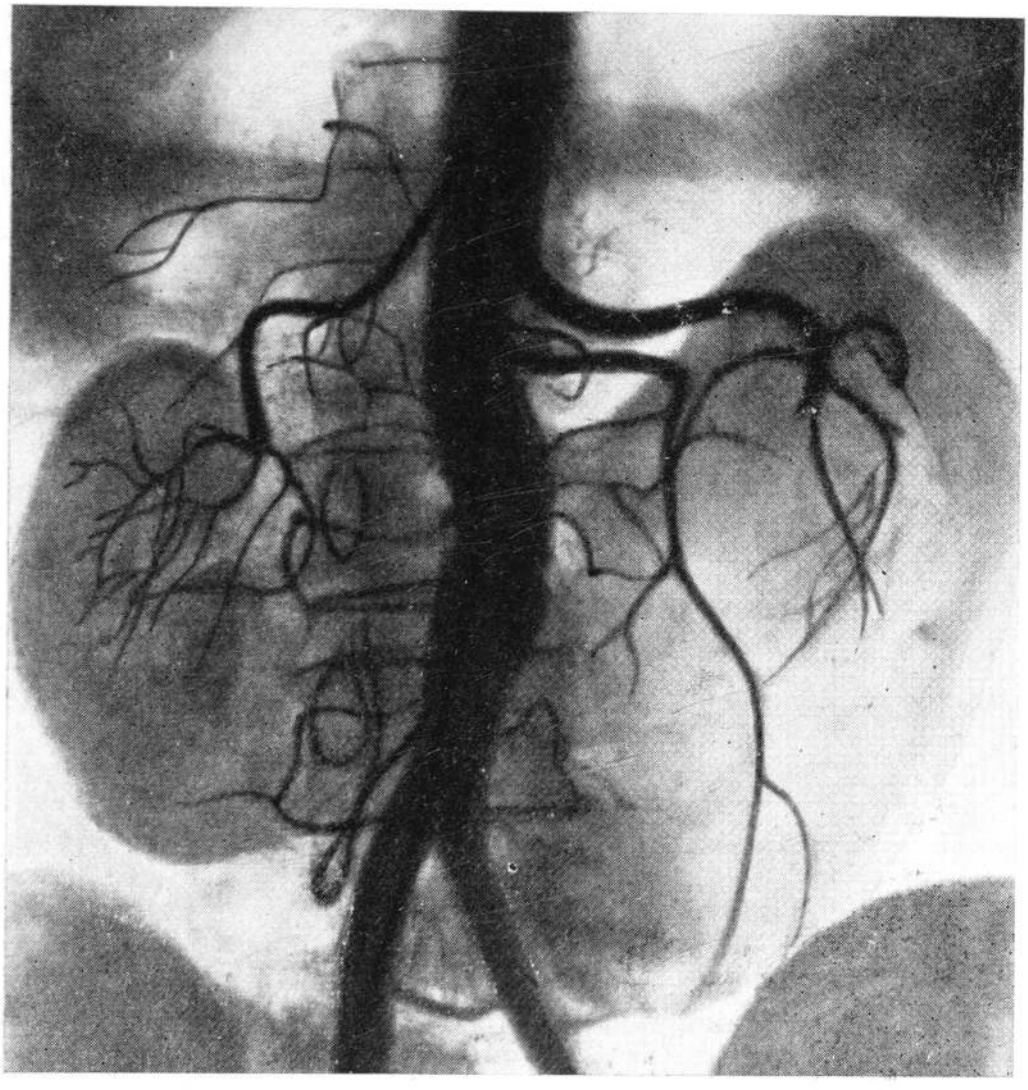


Fig. 45. Renal arteriography. Arterial phase. Horseshoe kidney. Tumour in its left half

tribution of the vessels in it the surgeon can avoid excessive and dangerous bleeding. The number and course of the vessels are determined in the arterial phase (Fig. 45), which indirectly gives an idea of the morphological structure of the kidney. The shape and position of the organ are determined in the parenchymatous phase (Fig. 46). The amount of contrast medium in the urinary tract in the final phase of the examination characterized the function of the kidney.

Pyeloscopy helps in distinguishing fused kidneys from bilateral renal dystopia by the different displacement of the kidneys during the examination.

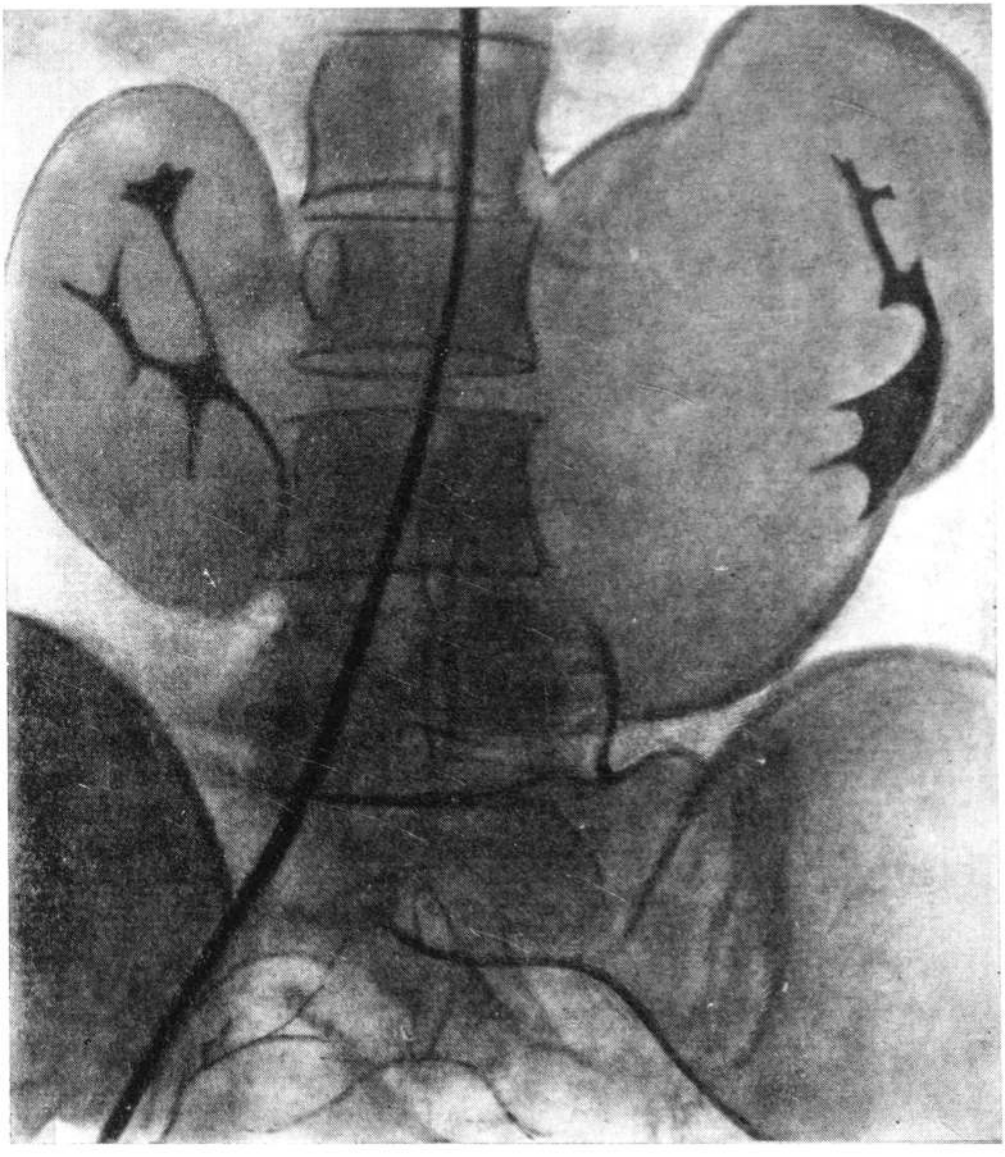


Fig. 46. Renal arteriography. Parenchymal phase. Horseshoe kidney. Tumour in its left half

Judging from our cases, **urokymography** makes it possible to determine precisely the tonus of the upper urinary tract and any latent disorders of its function. In cases of fused kidneys the urokymograms revealed lively, rhythmic and physiologically oriented contractions of the pelves and ureters, often with prolonged systolic or diastolic phases (Fig. 47). The depth of the pelvic waves was 15 to 33 mm, that of the ureteral waves 3 to 5 mm. The waves formed at intervals of 7 to 10 minutes at a rate of 2 to 3 cm/sec. Different degrees of lateral displacement

Fig. 47. Urokymograph. Horseshoe kidney. Dyskinesia of the upper urinary tract

of the prepelvic ureteral parts were noted in most cases. Sometimes the contractions of the upper third of the ureters in such cases were weaker.

Thus, in most cases in this group disorders of the motor function of the upper urinary tract were revealed, namely deeper

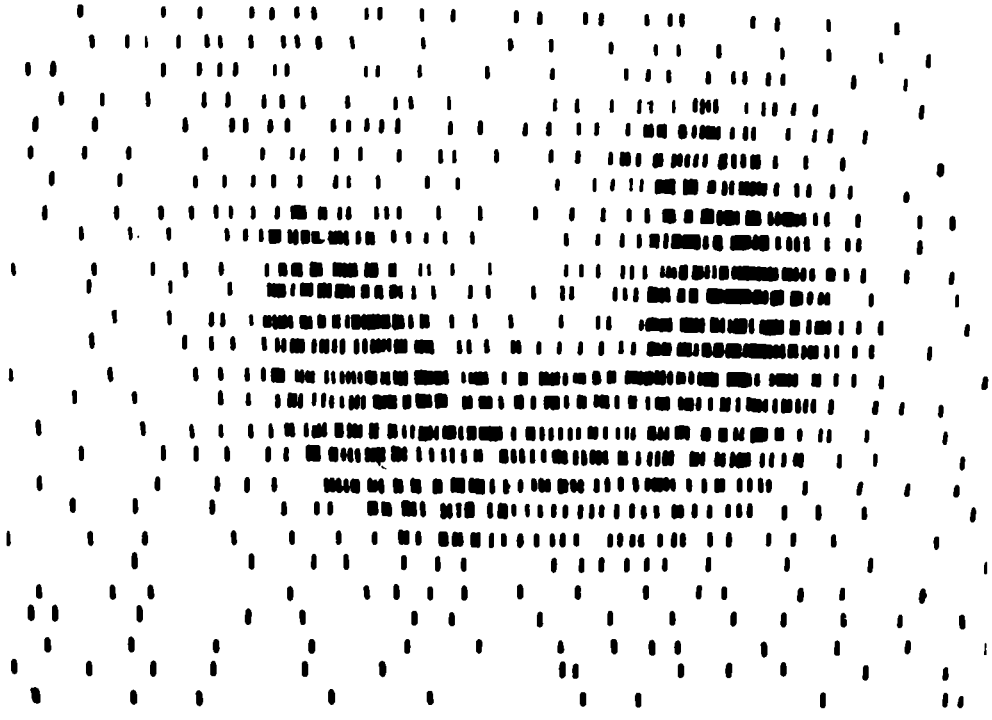


Fig. 48. Scintiscan. Horseshoe kidney

pelvic waves, changes in the indices of the systole-diastole complex, some increase in the rate of contractions, and lateral displacement of the prepelvic parts of the ureters above the isthmus or lower pole of the kidney. It should be emphasized that all these changes are associated with the pelvis or upper third of the ureter. Contractions in the other parts of the ureters were normal.

We encountered the above disorders of the upper urinary tract of fused kidneys, in the absence of evident diseases, only in those patients who had complaints, of pain in particular. This suggests that the patients' complaints are linked with the disorders. It may be assumed that compression of the nerve plexuses by the anomalous organ is not the only factor that gives rise to the patients' complaints. A definite role is played by compression of the prepelvic part of the ureter above the kidney and chronic stasis of the urine in the pelvis, which lead to disturbed dynamics of urine elimination from the pelvis and upper third of the ureter.

It is possible that an adhesive process, frequently found during operation, contributes to compression of the ureter above the kidney. Compression of the ureter during its contraction, however, also cannot be excluded. During contraction the ureter, which is curved forward, may tighten and come in contact

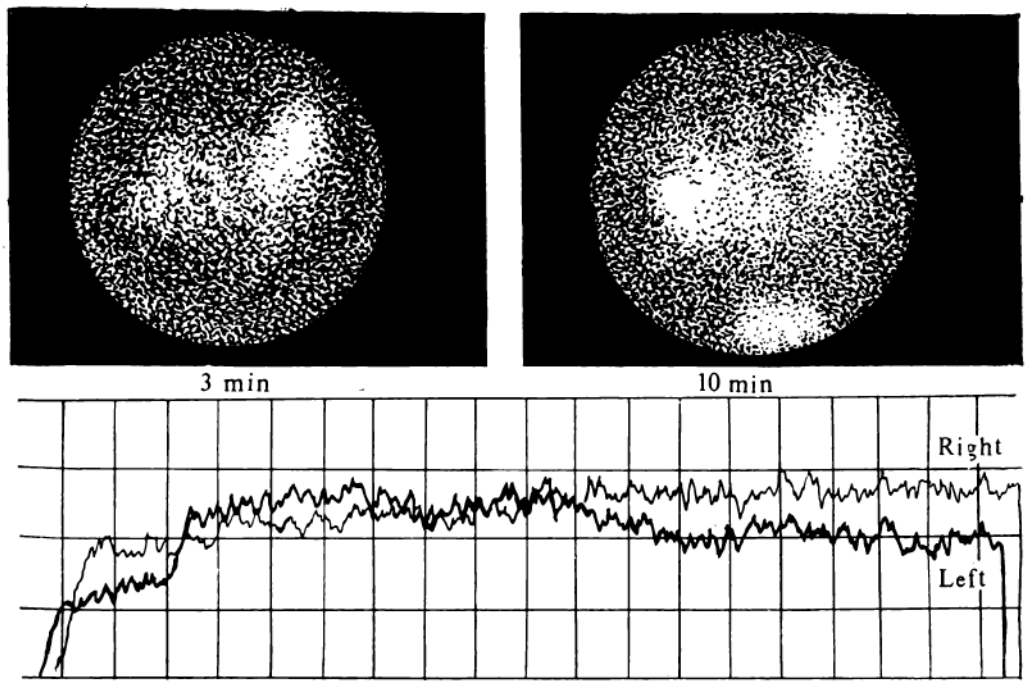


Fig. 49. Dynamic scintigraphy with renography. Horseshoe kidney

with the kidney, and this probably is the very cause of the lateral displacement of the ureter, the weakening of its contractions, and the disturbance of the systole-diastole complex. Impaired drainage of urine from the pelvis leads to high-amplitude contractions. The depth of the pelvic wave under normal conditions is 3 to 5 mm; in cases with fused kidneys it is usually 15 to 33 mm. The congenital character of these abnormalities is probably conducive to the unstable compensation, which is disturbed with time and with increased load, as the result of which a disease associated with the retention of urine in the pelvis develops. Thus, urokymography helps in revealing the early symptoms of the disease before marked morphological changes develop in the kidney. This is of considerable importance because indications for surgical management will be considered in time. Contrast pyelotelescopy and cineurography provide information on the dynamics of the upper urinary tract of a fused kidney.

Isotope renography and scanning of the kidneys may be valuable in the diagnosis of fused kidneys and study of their functional capacity. With this method a fused kidney is recognized by its direct image (Fig. 48). Scanning gives a general idea of the anatomical features, function, and morphological changes of the different parts of such a kidney. Its diagnostic value is less in diseases with diminished functioning of the kidney.

Horseshoe and unilateral fused kidneys produce a single shadow on the scintiscan, as distinct from two dystopic kidneys.

Examination with a gamma chamber produces clearer data on the anatomical and functional condition of a fused kidney (Fig. 49).

All the above-mentioned methods of examination have proved quite adequate in making the proper diagnosis of a fused kidney in our cases, in excluding the existence of disease, and in gaining a clear idea of the structure and function of the kidney before operation.

Errors in diagnosis and differential diagnosis. Despite the considerable success achieved in the diagnosis of different renal diseases, in many cases the diagnosis of fused kidneys is still made late.

In many cases fused kidneys were recognized at our clinic concurrently with the development of the disease. Only in one quarter of patients was the diagnosis made earlier, during surgical intervention or urological examination. In most cases with clinical manifestations the diagnosis was made 3 to 20 or more years after the appearance of the first symptoms.

Fused kidneys were often the cause of various diagnostic errors. The symptomatology of the renal anomaly was taken as signs of diseases of different organs. Patients underwent surgery for erroneously diagnosed appendicitis, cholecystitis, duodenal ulcer or diverticulum, ileus, gastric ulcer, pancreatic cyst, a disease of the liver, tuberculosis of the mesenteric lymph glands, retroperitoneal tumours or tumours of the intestine or new growths or cysts of the uterus and adnexa. During these operations an anomalous kidney was found to be the true cause of the pain. There were also errors of another kind. Tuberculosis of the mesenteric glands was, for example, taken for a fused kidney.

For many years our patients had been treated for other diseases which they did not actually have.

Diagnostic errors often led to unjustified operations. They were performed on 38 per cent of patients in this group: appendectomy, diagnostic laparotomy, and appendectomy and diagnostic laparotomy undertaken at various times after the first operation. The following case is given as an example.

T., a 16-year-old girl, was admitted to the clinic on February 18, 1959 complaining of a dull pain in the right abdomen and constipation. She claimed to have been ill for 12 months, during which time she had undergone appendectomy and, later, diagnostic laparotomy; the operations, however, did not bring relief. Postoperative scars were seen on the midline of the abdomen and in the right iliac region. An elastic, painless, and hardly mobile tumour was palpated to the right of the umbilicus. Blood and urine tests did not show any abnormalities. Excretory urography showed that both pelves were located at a low level and close to the vertebrae, and against

the shadow of a horseshoe kidney. The calyces were directed toward the vertebrae. A horseshoe kidney was diagnosed.

This case shows that such unjustified operations do not always reveal the proper diagnosis, though the anomaly can be recognized by means of excretory urography, which can be conducted in any surgical department. Surgery would not have been necessary if the examination had been done previously. Failure to resort to urological examination explains the occurrence of most diagnostic errors. The fact that a wide circle of physicians is insufficiently acquainted with this renal anomaly is of no little significance.

In most cases the possibility of a fused kidney should be obvious, and it should not be difficult to rule out diseases in the kidney or diseases of other organs, if the symptoms discussed above are found and a proper interpretation of radiographs taken during excretory urography or retrograde pyelography is made.

Sometimes the information obtained by excretory urography and retrograde pyelography is insufficiently convincing for the differentiation of fused kidneys from bilateral kidney dystopy, bilateral nephroptosis, or from some diseases involving the rotation and displacement of the kidneys, especially with atypical X-ray pictures of the upper urinary tract. In such cases the diagnostic difficulties may be solved by means of data gained from pneumoretroperitoneum.

Differential diagnosis is particularly difficult in cases of fused kidneys and concurrent diseases of other organs. The diagnosis of fused kidney and the disease in another organ has to be verified and a disease of the fused kidney ruled out. Otherwise the clinical manifestations of the ailment of the other organ may be taken for renal disease. The following case illustrates this.

L., a 58-year-old woman, was admitted to the clinic on March 22, 1968. She complained of dull pain in the right iliac region, high body temperature, and restriction of movement in the right lower limb. She had earlier been treated at a surgical clinic where diagnostic laparotomy revealed a horseshoe kidney. The disease was attributed to it. On admission to the urological department sharp pain was noted in the right ilio-inguinal region. The right lower limb was adducted and extension at the hip joint was very painful. An elastic, painless, and poorly mobile tumour was palpated below the umbilicus. Blood test: Hb 77 per cent, leucocytes 19 000, eosinophils 1, rod neutrophils 5, segmented neutrophils 71, lymphocytes 17, monocytes 6, ESR 58 mm/hour. Urinalysis showed normal values. The results of retrograde pyelography verified the diagnosis of horseshoe kidney and helped in ruling out any disease in it. Paracolic abscess was suspected. The diagnosis was confirmed during operation. The abscess was opened and a drain introduced into it. No connection between the abscess and the kidney was found.

In this case laparotomy assisted in the recognition of the renal anomaly, but the signs of paracolic abscess were taken for

symptoms of this developmental anomaly. Urological examination provided the proper diagnosis.

Sometimes the incorrect interpretation of the clinical manifestations of a fused kidney leads to the disclosure of a latent ailment of some other organ, as was the case with the following patient.

K., a 44-year-old woman, was admitted to the clinic on April 23, 1968. She complained that for the last three years she had experienced recurrent dull pain in the epigastrium. Appendectomy had been performed for that reason but the pain persisted. The stomach was later examined and a polyp found. Gastric resection also failed to relieve the pain. The patient was then examined in a urological clinic. Postoperative scars were seen in the right iliac region and on the midline above the umbilicus. Blood and urine tests showed normal values. An image of a horseshoe kidney in the lumbar region was produced on a scout radiograph. Excretory urography showed that the upper urinary tract was in a position characteristic of this anomaly. This examination verified the diagnosis of a horseshoe kidney without any doubt.

In this case, the clinical manifestations of the horseshoe kidney were apparently attributed first to appendicitis and then to polyp of the stomach. They persisted after removal of the vermiform appendix and after resection of the stomach.

Timely recognition of fused kidneys is very important because diagnostic errors and unjustified operations will thus be avoided and proper treatment prescribed.

PECULIARITIES OF THE SYMPTOMATOLOGY, CLINICAL COURSE AND RECOGNITION OF DISEASES OF FUSED KIDNEYS

Hydronephrosis

The anatomical features of fused kidneys predispose the individual to hydronephrosis. Our practice shows it to be the most common in this developmental anomaly of the kidneys. We encountered it in 34.3 per cent of our patients.

The particular predisposition of fused kidneys to retention ailments is conducive to bilateral lesions. We observed unilateral hydronephrosis in only 37 per cent of cases, and bilateral in 63 per cent. In two cases of unilateral hydronephrosis the contralateral half of the kidney was affected with nephrolithiasis and pyelonephritis.

It should be noted that the disease is encountered with equal frequency on each side. In 53 per cent of our patients the right half of the fused kidney was hydronephrotic, and in 47 per cent the left half.

The above-noted causes also promote the development of more

than one disease in one half of the kidney. In 78 per cent of our cases the hydronephrotic kidney was affected with pyelonephritis (which was bilateral in nine patients), and in 9 per cent with nephrolithiasis (bilateral in one patient). Besides, one patient had trauma of the hydronephrotic kidney.

In hydronephrosis of fused kidneys there are several factors which impair drainage of the urine from the pelvis: the high origin of the ureter from the pelvis, as a result of which a 'spur' or valve forms; the ureter passing over the kidney or its isthmus; the ureter branching off from the pelvis at a sharp angle; the prepelvic part of the ureter being attached to the isthmus of the kidney by means of periureteral adhesions; fixated ureteral kinks; ureteral strictures; stenosing periureteritis; compression of the prepelvic part of the ureter by accessory vessels; vesicoureteral reflux; nephrolithiasis. The frequency of the different causes of hydronephrosis is shown in Table 3 (only cases in which the cause was determined were taken into account).

Table 3. Causes of Hydronephrosis of Fused Kidneys

Cause of hydronephrosis	Number of cases
High origin of ureter +	
(a) compression on renal isthmus	14
(b) compression on renal isthmus + nephrolithiasis	2
(c) accessory vessel + compression on renal isthmus	3
(d) accessory vessel + nephrolithiasis	1
(e) fixated ureteral kink	2
(f) fixated ureteral kink + compression on renal isthmus + nephrolithiasis	1
(g) stricture of pelviureteral segment	3
(h) stricture of pelviureteral segment + compression on renal isthmus	2
(i) stricture of pelviureteral segment + compression on renal isthmus + nephrolithiasis	1
(j) stenosing periureteritis + compression on renal isthmus	5
(k) stenosing periureteritis + nephrolithiasis	1
Accessory vessel + compression on renal isthmus	2
Stricture of pelviureteral segment	1
Stenosing periureteritis +	
(a) compression on renal isthmus	5
(b) compression on renal isthmus + vesicoureteral reflux	1
Total :	44

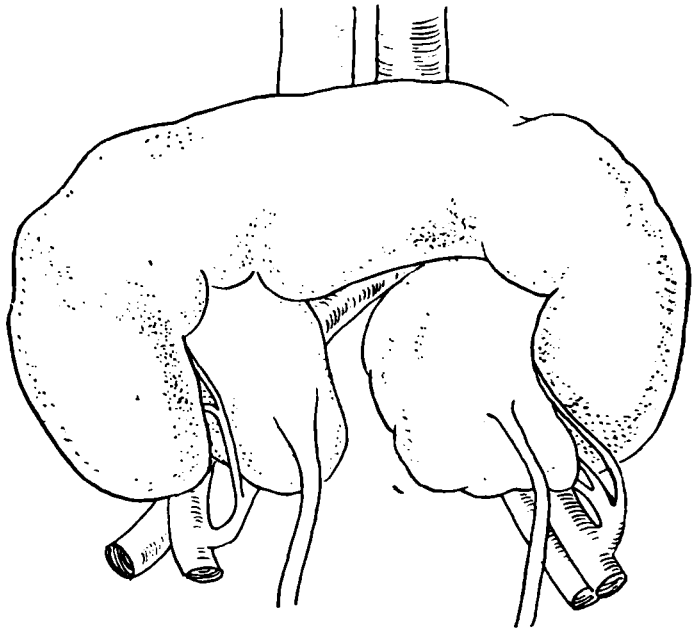


Fig. 50. Bilateral hydronephrosis of horseshoe kidney fused at the upper poles (from postmortem examination)

According to our experience the most common cause of hydronephrosis in cases of fused kidneys is high origin of the ureter, although it is often concurrent with other causes.

With the high origin of the ureter there are unfavourable conditions for the drainage of urine due to the considerable pressure in the lower part of the pelvis. Drainage of urine from the pelvis is particularly impaired because of the characteristic anterior position of the fused kidney. In many cases the ureters arise from the pelvis at a sharp angle. Sudden increase in diuresis promotes forward displacement of the enlarged pelvis, because the liver is behind it. This compresses the ureter on the anterior surface of the pelvis ever more, and a 'spur' or a valve forms. The enlarged pelvis disturbs the circulation of the blood in the kidney and induces its final blocking, which occurrence is more frequent than in normal kidneys. The condition is also promoted by periureteritis developing due to the infection which is so frequently encountered in such kidneys.

High origin of the ureter is rarely encountered in fusion of the kidneys at the upper poles (Fig. 50).

The conditions are favourable for the fixation of the prepelvic part of the ureter stretching over the kidney. Attachment of the ureter to the isthmus or lower pole of the fused kidney by periureteral adhesions may itself be a cause of poor pelvic drainage.

It prevents contraction of this part and causes compression of the ureter.

The other causes of hydronephrosis are much rarer: fixated kink of the prepelvic part of the ureter was encountered in three cases, stricture of the pelviureteral segment in seven, compression of the ureter by an accessory vessel in six, vesicoureteral reflux in one, and nephrolithiasis in six cases.

The **symptomatology** of hydronephrosis of fused kidneys has certain peculiarities.

Pain of various intensity is a frequent symptom. We encountered it in all cases. Pain may be dull and continuous or it may occur in attacks. In some of our patients it had the character of renal colic. Such attacks occurred at different intervals ranging from several days to several months. The pain is often linked with physical exertion or a sudden change in the position of the body, which is characteristic of this kidney anomaly. Pain is not always felt in the lumbar region, patients often complaining of pain in the abdomen, hypochondrium, iliac or epigastric region. With such atypical localization of pain the clinical picture may resemble that of diseases of the abdominal organs.

In one of our patients the unusual localization of pain in the abdomen simulated the picture of ileus. In a disease of many years' duration this suggests chronic ileus, while in diseases of an acute character such pain may be interpreted as acute ileus.

It should be borne in mind, however, that compression of the duodenum between the mesenteric vessels and the horseshoe kidney isthmus, which is enlarged in stasis of the urine and circulatory disorders, may result in high arteriomesenteric occlusion. The following case is given as an illustration.

V., a 44-year-old man, was admitted to the surgical department on June 6, 1962 as an emergency case. He complained of severe spasmodic abdominal pain and frequent vomiting, which were relieved in a knee-elbow position. The abdomen was distended, and palpation revealed tenderness in different parts of it. High arteriomesenteric ileus was suspected. Diagnostic laparotomy showed that the ailment was caused by a horseshoe kidney. Pain subsided after the operation and the kidneys were examined in the urological department. An elastic, painless, and slightly mobile tumour was palpated on the left and below the umbilicus. Blood and urine tests showed normal values. Indigo carmine was discharged from the left ureteral orifice in the fifth minute and from the right in the sixth minute. Retrograde pyelography showed the pelves at the level of the second and third lumbar vertebrae, with the calyces directed medially. A diagnosis of bilateral hydronephrosis of a horseshoe kidney was made.

The atypical clinical picture of the disease is also determined by the abnormally low localization of the tumour in the abdomen; this was the case in 63 per cent of our patients with fused kidneys. The tumour is usually close to the vertebrae; sometimes it

is found in the hypochondrium. It is elastic, painless, and only slightly mobile. Sometimes the tumour lies across the spine and is shaped like an elevation with the ends pointing toward the hypochondria. The pulse of the vessels is felt above and below the tumour.

In some cases the tumour appears, disappears, and then reappears, which is characteristic of hydronephrosis. Its appearance is often attended by elevation of body temperature, a chill and positive Pasternatskii's sign. With the disappearance of the tumour, these symptoms also disappear. Polyuria develops at that time.

Patients in this group sometimes have symptoms which are considered characteristic of fused kidneys. For instance, Rovsing's and Gregoir's symptoms, cardiovascular disorders, neurological disturbances, and dyspeptic phenomena were noted in two to twelve cases. Since these symptoms are not specific and occur rarely they cannot be used in the diagnosis of renal anomaly.

In many cases of fused kidneys the clinical picture comprises the symptoms of the disease, but sometimes it consists only of the symptoms of the kidney anomaly. The following case illustrates this.

B., a 25-year-old woman, was admitted to the clinic on October 3, 1957. She complained of a dull pain experienced now and again in the region of the umbilicus during sudden changes in the position of the body, and constipation. An elastic, painless and hardly mobile tumour was palpated below the umbilicus, and vascular pulsation was felt above and below it. During exacerbation of the pain Pasternatskii's symptom was mildly positive on the right. Blood and urinary tests did not show any abnormalities. Indigo carmine was discharged from both ureteral orifices in the sixth minute. Excretory urography showed that both pelves were distended and located in the vicinity of the third to fourth lumbar vertebrae, their calyces pointing toward the midline. Diagnostic pneumoretroperitoneum allowed an image of a horseshoe kidney to be seen. A diagnosis of bilateral hydronephrosis of a horseshoe kidney was made.

Hydronephrosis of a fused kidney may follow an asymptomatic course.

Thus, according to the clinical picture four groups of patients with hydronephrosis of fused kidneys can be distinguished: (1) asymptomatic group; (2) a group with symptoms of the disease; (3) a group with symptoms of the disease and renal anomaly; (4) a group with symptoms of the renal anomaly. The clinical picture is often atypical and resembles that of diseases of the abdominal organs.

The clinical course of hydronephrosis of fused kidneys is usually chronic in character. We encountered an acute and subacute

course in only three patients. It may be assumed that this was the onset of the disease.

In some patients the first signs of the disease develop in childhood or in the first months of life, but often the ailment begins troubling the patient in later years. In most of our cases the duration of the clear period ranged from 20 to 40 years.

Often the disease follows a course of many years. The period from the time of the first symptoms to the time the patient is admitted to the hospital varies in most patients from three to twenty years. Because of the atypical clinical picture, patients are usually admitted to the clinic with marked hydronephrosis. Our cases, for instance, were distributed according to the stage of hydronephrosis as follows; 8 per cent stage I, 67 per cent stage II, and 25 per cent stage III.

Permanent dull pain is noted only in some cases. The clinical manifestations usually develop in attacks which are followed by clear periods, sometimes of one to two years. With each attack, however, the pain is more intense and the duration of the attack itself longer so that the patient finally becomes incapable of working. The course of the disease is particularly aggravated in frequent bilateral affection, which is conducive to the development of renal insufficiency.

The **diagnosis** of hydronephrosis of a fused kidney comprises recognition of the disease and recognition of the renal anomaly.

General clinical methods of examination enable one in some cases to suspect the disease or the developmental anomaly of the kidney. Six of our patients knew that they had an anomalous kidney before they came to the clinic. They had earlier undergone urological examination or operation. In some patients hydronephrosis could be suspected because of an abdominal tumour which would now appear, then disappear, and which was associated with renal symptoms. This tumour was close to the midline in most cases, which suggested a fused kidney. The remaining patients were admitted with symptoms of an ailment of the organs of the urinary system which called for urological examination.

The principal role in the diagnosis of hydronephrosis of a fused kidney is played by X-ray examination and the other diagnostic methods described above.

Excretory urography suggests the diagnosis of the anomaly and the disease in only some cases, mainly when infusion techniques are used.

When renal function is reduced or is absent due to hydronephrosis, it is impossible to make a proper diagnosis of the kidney anomaly and disease by means of excretory urography. From the anomalous shape and position of the upper urinary tract of

the other functioning half, dystopia of the kidney may be suspected. It is especially difficult to even suspect a kidney anomaly when the single functioning half of the kidney has a normal pelvis with the calyces facing laterally.

Retrograde pyelography is necessary for making a proper diagnosis of the disease as well as for verifying or ruling out the diagnosis of renal anomaly in doubtful cases. The examination is particularly advisable in cases where infusion urography is ineffective because of sharply diminished function of the kidney.

With this examination the diagnosis of renal malformation is based on the signs described above, i.e. low position of the pelves which are situated close to the vertebrae, medial direction of the calyces, a small basic angle of the 'lumbar triangle', etc. The diagnosis of a fused kidney cannot be made from the pyelogram when the hydronephrosis cavity is partly filled with the contrast medium.

It should be noted that the results of bilateral examination are necessary for reaching a correct diagnosis. But even this examination will not verify the diagnosis of the anomaly with assurance in cases of marked bilateral distension of the pelves and calyces, localization of these structures at a great distance from each other, and when the calyces are directed laterally.

Pyelograms taken at an angle of 45 degrees are very demonstrative. They can show stenosis of the prepelvic part of the ureter, its displacement to the front, and the ureter arising at a high level.

Thus, infusion urography and retrograde pyelography yield valuable information for the diagnosis of hydronephrosis and kidney malformation. However, they supply indirect evidence of a fused kidney, owing to which auxiliary methods of examination have to be resorted to in some cases.

Pneumoretroperitoneum is the most exact method in the diagnosis of hydronephrosis of fused kidneys. The pneumograms usually demonstrate clearly the shape of the anomalous organ and its components, the size of each half of the kidney and its isthmus, and the position of each half in relation to the other, the spine and other organs.

The degree of preservation of the renal tissue may be judged from the density of the shadow. With a sclerotic process in the perirenal fat half the fused kidney is sometimes seen on the radiographs. In such cases it is shaped like a horn whose base is superimposed on the shadows of the vertebrae. This sign is sufficient proof to make a diagnosis of horseshoe kidney.

The combination of pneumoretroperitoneum and excretory urography conveys valuable information. The image of the fused organ and the relationship of the distended upper urinary tract

may be seen on the radiographs and the function of the anomalous kidney can be judged.

Diagnostic pneumoretroperitoneum carried out with the patient lying on his back produces an insufficiently clear image of the isthmus of the fused kidney because it is hidden by the shadow of the vertebrae. For a better demonstration of the area of fusion, it is advisable to examine the patient at an angle of 45 degrees. On such pneumograms the kidney appears flatter and more elongated and its lower poles are usually deviated to the front. The image of the isthmus of a horseshoe kidney may also be produced by combining pneumoretroperitoneum with tomography.

Renal angiography is a valuable auxiliary examination in cases of hydronephrosis of fused kidneys. The diagnosis of hydronephrosis may be made when the picture shows few vessels and those that are seen are thin and tortuous, while the anomaly is recognized by the image of the malformed kidney in the nephrographic phase. This examination also helps in judging the function of the kidney. Renal angiography is to be recommended before surgery. It helps in determining the course of the main and accessory vessels and the renal areas which they supply with blood. However, the methods of examination described above usually enable the physician to make a proper diagnosis, so that angiography is rarely used.

Urokymography gives an idea of the function of the upper urinary tract of an anomalous kidney. The radiographs show a weakening or the absence of pelvic contractions, arrhythmia and reflux. In tonogenous dilatation the amplitude of the waves is high, while in myogenous dilatation it is low, reaching 1 or 3 mm, or non-existent. Besides, there are marked disorders of the systole-diastole complex.

Urokymography also helps in revealing some of the causes of hydronephrosis. With compression of the ureter above the renal isthmus there are no contractions of its prepelvic part and the ureter is displaced laterally, with stricture of the prepelvic part there is a break in the shadow of the contrast medium; with compression of the ureter by an accessory vessel the lower border of the pelvis is seen as an even line; with reflux we see contractions with a reverse wave.

The dynamics of duplicated urinary tracts of a fused kidney have some peculiar features. Where the ureters come in contact the systolic contractions of one ureter are superimposed on the diastolic contractions of the other, which changes the indices of the systole-diastole complex.

Thus, in many cases the diagnosis of hydronephrosis of fused kidneys presents considerable difficulties, mainly associated with the atypical clinical picture of the anomaly. That is why about

one third of our patients in this group were admitted with a diagnosis of hydronephrosis, while the remaining patients were sent for treatment with an erroneous diagnosis (nephrolithiasis, nephroptosis, appendicitis, ileus, etc.). For the same reason it took a long time to diagnose the anomaly in this group. These diagnostic errors could have been corrected by early urological examination.

Nephrolithiasis

Nephrolithiasis is encountered quite frequently in cases of fused kidneys. In our practice we observed it in 20.3 per cent of cases.

In 12 per cent of our patients nephrolithiasis was bilateral. If we consider each half of the fused kidney separately, then stones were found on the right side in 53 per cent and on the left in 47 per cent of cases. They were located in the kidney in 92 per cent of cases, in the kidney and ureter in 2 per cent, and in the ureter in 6 per cent. In most cases there was a solitary stone; in 32 per cent of cases the stones were multiple, and in 6 per cent recurrent.

Nephrolithiasis of a fused kidney is very often attended by infection. In the half of the fused kidney with the stone, pyelonephritis was noted in 97 per cent of our cases and hydronephrosis in 12 per cent.

One of the causes of this disease in fused kidneys may be obstruction of urine drainage in the distal parts of the urinary tracts. In most cases, however, no abnormalities are found in these parts of the urinary tracts, and nephrolithiasis in fact developed due to impaired drainage of urine from the pelvis in the upper third of the ureter and in its prepelvic portion. We want to emphasize that in fused kidneys nephrolithiasis is very often linked with impaired pelvic drainage, but that other commonly encountered causes of the disease are also important.

The **symptomatology** of nephrolithiasis of fused kidneys is also marked by some specific features. All our patients in this group had pain, which in most was of the character of renal colic. Pain was encountered in this disease more often than in non-complicated anomaly or in other diseases. Dull pain persisted in the periods between attacks. Quite often pain occurred after physical exertion, or when the person was riding on a horse or in a car. It was felt in the small of the back or in the hypochondrium, but often it was located in the epigastrium, the iliac or umbilical region, or experienced throughout the whole abdomen and thus resembled pain encountered in diseases of the abdomi-

nal organs. This can be explained by the low position of the malformed organ.

A tumour palpated in the abdomen is a very frequent symptom. We found it in half of our patients in this group. This elastic and hardly mobile tumour, which may be painless or mildly tender, is usually located close to the vertebrae. Sometimes it is felt as a swelling lying across the spine.

According to the clinical picture, two groups of patients with nephrolithiasis of fused kidney can be distinguished: one with symptoms of the disease and the other with symptoms of the disease and the kidney anomaly.

Symptoms of nephrolithiasis were found in many patients in this group.

Rarely, in one to five of these cases, symptoms of the renal anomaly were added to the clinical picture of nephrolithiasis. The more frequently encountered dyspeptic signs are usually associated with renal colic.

Thus, nephrolithiasis of a fused kidney sometimes has an atypical clinical picture with pain and a tumour located in the abdomen close to the vertebrae. This last symptom should indicate to the physician the possibility of a fused kidney. In most cases the clinical picture is that of the disease. Symptoms of the kidney anomaly are rare and help little in the diagnosis.

The **clinical course** of nephrolithiasis of a fused kidney is chronic in character with periods of exacerbation. In our patients the first symptoms of the disease developed in childhood or at the age of 20 to 60. The clear period in most of them ranged from 10 to 40 years. Most patients had had the disease for many years. Three to 20 years had elapsed from the appearance of the first symptoms of the disease to the time of admission to our clinic. It is possible that some of the patients were first troubled by the symptoms of the renal anomaly.

The appearance of the symptoms of the disease after those of the anomaly often makes the patient think that the disease had developed in childhood, as in the following case.

B., a 7-year-old boy, was admitted to the clinic on July 14, 1958. He complained of sharp pain in the right lumbar region and right hypochondrium, which was attended with vomiting. He had been ill for six years. At first he had a dull pain in the abdomen. During the last year it became localized in the right half of the abdomen and occurred in attacks. One month before admission to the urological clinic he underwent appendectomy, but it did not bring relief from the pain. On inspection a postoperative scar was noted in the right iliac region. Pasternatskii's sign was positive on the right. Blood test: Hb 72 per cent; leucocyte count 7100; ESR 35 mm/hour. Urinalysis: reaction acid; specific gravity 1020; protein 3.3 pro mille; leucocytes covering the field of vision; erythrocytes 20-30 in field of vision. Urine culture produced growth of *Staphylococcus albus*. The outlines of a horseshoe kidney could be traced on the scout radiograph, on the right

of the third lumbar vertebra the shadow of a concrement (1.5×1.5 cm) was seen. Excretory urography showed that both pelves were located near the second and third lumbar vertebrae, on the left the calyces were directed medially. The diagnosis of stone and pyelonephritis of the right half of a horseshoe kidney was made. Pyelolithotomy was performed.

In the other patients of our group the onset of the disease was marked by acute symptoms and the clinical picture was that of nephrolithiasis. This suggests that before the disease the renal anomaly was asymptomatic.

Thus, nephrolithiasis of fused kidneys is a chronic disease which is usually characterized by progressive development of the symptoms and pathological changes in the kidney. A bilateral process leads to renal insufficiency.

The **diagnosis** of nephrolithiasis of fused kidneys is sometimes very difficult, it being especially difficult to recognize the kidney anomaly. A fused kidney may be suspected in cases when a painless and hardly mobile tumor is palpated in the abdomen on the midline. In the remaining cases a general clinical examination and laboratory tests yield data for making a diagnosis of urolithiasis and suggesting urological examination.

Scout X-ray of the urinary tract made it possible to recognize the disease in almost all of our cases. In only one case could the stone not be seen on the radiograph.

Scout X-ray is of great help when the image of a horseshoe kidney is demonstrated on the radiograph. We found this sign in half of the patients examined.

A scout radiograph showing stone shadows located at a low level and close to or very far away from the vertebrae suggests their localization in an anomalous kidney. In those rare instances when the branches of the coral concrement are directed medially, or when numerous stones occupy both halves of the kidney or one half but follow the shape of the anomalous pelvis a more definite opinion on the presence of a fused kidney may be formed on the basis of the shape and position of the stones on the scout radiograph. Even in these cases, however, contrast methods of X-ray examination have to be used to identify the type of kidney anomaly.

Excretory urography may sometimes, when renal function is maintained, yield sufficient information for making a diagnosis of fused kidney and nephrolithiasis. In such cases both pelves are situated at a low level and close to the vertebrae on the radiographs while their calyces are directed medially and are included in the 'lumbar rectangle'. The stones contribute to the retention of the contrast medium and the upper urinary tracts are demonstrated quite clearly on such radiographs (Fig. 51).

These radiographs are especially useful when they show the

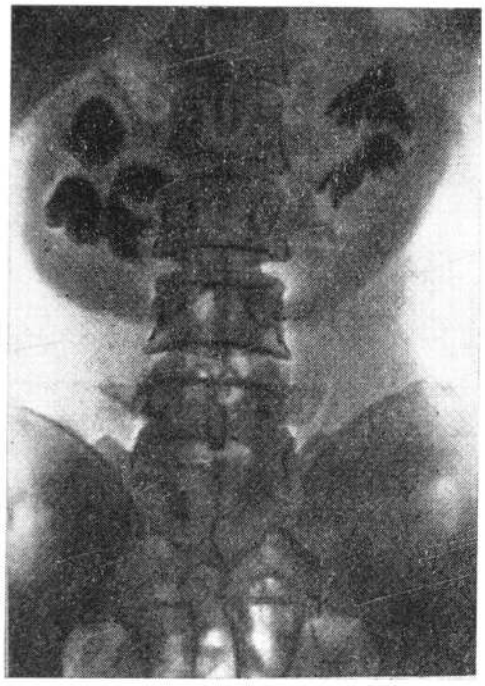


Fig. 51. Excretory urography. Stones in both halves of a horseshoe kidney

image of a fused organ. There is no doubt then about the diagnosis of a fused kidney. Besides, the anatomical and functional changes in the kidney may be judged from this examination.

Even with good renal function the results of excretory urography are not always sufficient for diagnosing a fused kidney, and when renal function is reduced the diagnostic possibilities of this method are even more limited. In such cases, the anomaly should be suspected when the image of an anomalous pelvis is seen on one side and distended calyces medial of the pelvis on the other. Sometimes the radiographs show distended calyces situated close to the vertebrae and all produce the image of half of a horseshoe kidney while the contralateral pelvis has an anomalous shape. In some cases the diagnosis of a horseshoe kidney can be made both from the position of the stones characteristic of this anomaly and from the malformed pelvis.

In cases in which the radiograph does not show the image of one of the pelves and the stones are not in the position characteristic of the renal anomaly then the image of a single malformed pelvis may suggest unilateral dystopia of the kidney. The stones are usually seen on the radiograph on the level of or below the other malformed pelvis.

It is very difficult to suppose that the patient has a fused kidney when the pelves are far away from each other and the calyces are directed laterally. In such cases the proper diagnosis may be made by means of auxiliary examination: a palpated

tumour in the region of the umbilicus, the image of a fused kidney on scout radiographs, the findings of diagnostic pneumoretroperitoneum.

Retrograde pyelography is a valuable method in the diagnosis of nephrolithiasis of fused kidneys. The radiographs often show the signs described above as characteristic of a fused kidney.

As is common in nephrolithiasis, the examination has to be resorted to in the diagnosis of roentgen negative stone and when the kidney is not functioning.

Pneumoretroperitoneum is used, but rarely, in the diagnosis of nephrolithiasis in a fused kidney. It demonstrates the signs described above.

In nephrolithiasis the gas sometimes does not penetrate the retroperitoneal space due to sclerotic paranephritis. In such cases the anomaly may be recognized by the image of half the horseshoe kidney seen as a 'horn'. Sometimes the isthmus of the fused organs is not clear on the radiographs, but the shape of the lateral lobes is indicative of the renal anomaly. Pneumography conducted at an angle of 45 degrees helps in making an exact diagnosis in such cases. The radiographs show the image of an elongated kidney whose inferior pole is in front of the vertebrae and is continuous with the pole of the other kidney.

Urokymography in nephrolithiasis of fused kidneys shows weakening of the contracted pelvis with waves of very low amplitude or stenotic contractions with large-amplitude waves, which correspond to those in myogenous or tonogenous dilatation of the pelvis in hydronephrosis. The contractions of the prepelvic part of the ureter are usually arrythmical and weak. Disturbance of the systole-diastole complex and lateral displacement are noted in this segment of the ureter. If there is no stone, the motor function of the middle and lower third of the ureter is usually normal.

The methods of examination described above provide the proper diagnosis of the kidney anomaly and disease in all cases. Diagnostic errors occur when urological examination is not undertaken.

Pyelonephritis

Pyelonephritis often accompanies nephrolithiasis and hydronephrosis of fused kidneys, but may also be encountered as an independent disease. Among our patients 7.0 per cent had pyelonephritis as the principal ailment; it was bilateral in most of them.

Urinary stasis, with all its ensuing consequences, contributes greatly to the development of this disease in a horseshoe kidney.

The **symptomatology** of pyelonephritis in a fused kidney differs rather from that of the disease in a normal kidney. Two groups of patients can be distinguished according to the clinical picture: a group with symptoms of the disease, and a group with symptoms of the disease and of the renal anomaly.

The common complaint is pain experienced now and again in the small of the back or in the hypochondrium. Sometimes it is felt in the region of the umbilicus and the epigastrium close to the midline. The pain is dull and rarely of a sharp character.

In many patients a tumour characteristic of a horseshoe kidney is palpated in the abdomen close to the midline; it is elastic, painless, and hardly mobile. The uncommon localization of the pain and the tumour causes considerable diagnostic difficulties.

In most cases the clinical picture is that of pyelonephritis.

The symptoms of the renal anomaly are very rarely encountered. Only one of our patients in this group had a positive Rovsing's sign, and two had dyspeptic complaints.

The **clinical course** of pyelonephritis of fused kidneys is chronic with periodical exacerbations. The disease is usually displayed at a mature age. In our patients the duration of the clear period ranged from 10 to 40 years. Once developed, the disease lasts years. A period of 2 to 10 years had elapsed from the appearance of the first symptoms to admission of our patients to the clinic.

The **diagnosis** of pyelonephritis in these patients is usually made without any difficulty using commonly accepted methods of examination. Patients are sent to the clinic for examination because of the disease, while the anomaly remains unrecognized.

In this disease also X-ray examination plays the main role in the diagnosis of the fused kidneys. The anomaly is revealed by the methods described above.

Tuberculosis

Tuberculosis of fused kidneys is a rare occurrence. We encountered it in 6.2 per cent of our patients. It is usually found in association with other diseases because of the anatomical features of the upper urinary tract. One of our patients had tuberculosis of the kidney, hydronephrosis, pyelonephritis and nephrolithiasis.

The medical history of these patients is often marked by specific affection of other organs.

Symptomatology. According to the clinical picture, three groups of patients with tuberculosis of fused kidneys can be distinguished: an asymptomatic group, a group with symptoms of the disease, and a group with symptoms of the disease and the anomaly. The case described by Kusevitsky et al. (1961) illustrates an asymptomatic course of the disease. Tuberculosis of a fused kid-

ney was a chance finding on autopsy for gastric carcinoma.

The disease is manifested by different symptoms, the most frequent of which is pain. It is localized in the small of the back or the epigastrium and is usually dull, rarely sharp.

The uncommon localization of pain in the epigastrium creates considerable diagnostic difficulties. The following case is given as an illustration.

K., a 49-year-old man, was admitted to the clinic on September 2, 1968. He complained of dull epigastric pain, insomnia, irritability, fatigue, and constipation. He had had these symptoms for five years; in the last year he had also experienced dull pain in the left hypochondrium and, at times, frequent and painful micturition. Objective examination showed no abnormalities. Blood test produced normal values. Urinalysis: reaction acid; specific gravity 1016; protein 0.99 pro mille; leucocytes covering the entire field of vision; erythrocytes 15-20 in field of vision. The floatation method demonstrated *Mycobacterium tuberculosis* in the urine. Indigo carmine was discharged from the right ureteral orifice in the 5th minute and from the left orifice in the 7th minute. A scout urogram showed the shadow of a horseshoe kidney. Retrograde pyelography showed both pelves on a level with the second lumbar vertebra and close to it, the calyces directed medially, and the ureters in the upper third deviating laterally from the vertebrae. The inferior calyx of the left pelvis was distorted and typical of tuberculous papillitis.

The localization of pain in the epigastrium in this case was evidently associated with the clinical manifestations of the renal anomaly. With the development of the concomitant disease the painful sensations became more intense, and pain was also felt in the hypochondrium. Thus, the atypical picture of the disease was apparently due to the concurrence of the symptoms of the disease and those of the anomaly.

The clinical picture of the disease is also atypical when the tumour is found at an uncommon site, in the region of the umbilicus. We found such localization in five of eight patients in this group. Sometimes the elastic, painless, and hardly mobile tumour has the shape of the swelling characteristic of a horseshoe kidney and lies across the spine. This justified a high degree of suspicion of a fused kidney.

The clinical picture of tuberculosis of fused kidneys is sometimes complicated by symptoms of several concomitant diseases. The following case from our practice serves as an example.

P., a 41-year-old woman, was admitted to the clinic on September 19, 1965 complaining of permanent dull pain in the right lumbar region and frequent and painful micturition experienced for eleven years. In 1954 tuberculosis of the right kidney was diagnosed through urological examination and surgery was suggested, to which the patient would not agree. She received various antituberculotics at short intervals. In the last year her condition had become worse. Blood test: haemoglobin 72 per cent; leucocytes 5200: eosinophils 1, rod neutrophils 3, segmented neutrophils 51, lymphocytes

40, monocytes 5; ESR 29 mm/hour. Urinalysis: reaction acid; specific gravity 1014; protein 0.99 pro mille; leucocytes covering the entire field of vision; erythrocytes 30-35 in field of vision. Urine from the left kidney contained 2-3 leucocytes in field of vision. Urine culture produced growth of streptococcus. The floatation method demonstrated *Mycobacterium tuberculosis* in the urine. Cystoscopy revealed focal hyperaemia and miliary tubercles characteristic of tuberculosis at the right ureteral orifice. Indigo carmine was discharged from the left ureteral orifice in the fifth minute, while none was discharged from the right orifice within 12 minutes. A scout radiograph showed two round shadows of concrements, measuring 2×3 cm and at a distance from the vertebrae, in the right hypochondrium. Retrograde pyelography demonstrated a very large right pelvis with the calyces directed medially. Urograms showed two small pelves on the left of the twelfth thoracic and second lumbar vertebrae, and numerous contrast spots on the right at the same level close to the vertebrae. Tuberculo-infiltrative tuberculosis of the right half of a horseshoe kidney, hydronephrosis, pyelonephritis, and nephrolithiasis, duplication of the left half of the kidney with fissure of the ureter were diagnosed.

In this case symptoms of several diseases were noted, tuberculosis, hydronephrosis, pyelonephritis, and nephrolithiasis, as well as duplication of the left half of a horseshoe kidney.

In patients of this group the symptoms of the disease usually predominate over those of the kidney anomaly. Disorders of micturition, pyuria, haematuria, proteinuria, elevated body temperature, *Mycobacterium tuberculosis* in the urine, positive Pasternatskii's sign are frequent symptoms; some patients have arterial hypertension, focal hyperaemia and miliary tubercles on the mucous membrane of the urinary bladder, and changes in the differential blood count. Neurological changes and dyspeptic signs are noted occasionally.

The **clinical course** of tuberculosis of fused kidneys is chronic in character. The disease continues for many years due to the symptoms of the anomaly being later accompanied by the symptoms of tuberculosis.

The **diagnosis** of tuberculosis of the kidneys in these cases is quite easy. It is based on the history of the specific process in another organ, pyuria, identification of *Mycobacterium tuberculosis* in the urine, specific tubercles on the bladder mucosa, and destruction of the calyces, i.e. on the commonly encountered signs of the disease.

The difficulties are usually associated with the diagnosis of the kidney anomaly. A fused kidney is suspected when the characteristic tumour is palpated in the umbilical region and X-ray methods of examination yield the data described above. Retrograde pyelography is used most often in this disease to disclose the early X-ray signs of tuberculosis and differentiate them from those of the pyelo-calyceal system of fused kidneys.

Errors in the diagnosis of tuberculosis of fused kidneys are usually due to inadequate X-ray urological examination.

Nephroptosis

Nephroptosis rarely occurs as an independent disease in fused kidneys. We encountered it in only 3.1 per cent of our cases, which can be explained by the strong fixation of the fused organ by the isthmus above the vertebrae and the accessory vessels.

The **symptomatology** of nephroptosis of fused kidneys is usually characterized by the symptoms of the disease.

Examination in all cases reveals pain associated with muscular exertion, and a mobile tumour in the abdomen. The pain is usually felt in the abdomen but may sometimes be referred to the small of the back. In cases of horseshoe and cake kidneys the tumour is palpated on the midline, while in patients with unilateral fused kidneys an elongated tumour is found in the hypochondrium. In addition, dysuria and a positive Pasternatskii's sign may sometimes be observed. The uncommon localization of pain and tumour makes the recognition of the anomaly and disease difficult. The following case is given in illustration.

K., a 38-year-old man, was admitted to the clinic on April 19, 1958. He complained of attacks of pain experienced in the right iliac region over a period of three years. They occurred during work or when he rode in a car over an uneven road. One month before admission to the urological department he underwent appendectomy, but pain recurred after the operation. A fresh postoperative scar was seen in the right iliac region. An elastic, painless, and mobile tumour was palpated in the umbilical region. Hypospadias was also found. Blood and urine tests showed normal values. Indigo carmine was discharged from the orifices of both ureters in the fourth minute. An image of a horseshoe kidney was produced on a scout radiograph. Excretory urography demonstrated malformed renal pelves with the calyces facing the vertebrae against the background of an image of a horseshoe kidney. On examination, with the patient standing, the pyelo-calyceal system was displaced downward by 5 cm. A diagnosis of nephroptosis of a horseshoe kidney was made.

The atypical clinical picture in this case led to a diagnostic error and an unjustified operation.

Nephroptosis of fused kidneys follows a chronic clinical course. The disease usually develops at a mature age and lasts many years.

Diagnosis. This renal anomaly may be suspected on the basis of the characteristic clinical and X-ray symptoms mentioned above. The displacement of the kidney is revealed by methods commonly applied in nephroptosis.

Patients with nephroptosis of fused kidneys usually come to the clinic because of the disease, so that the renal anomaly is recognized at the time of the development of the disease.

New Growths of Kidney

New growths of fused kidneys are a rare occurrence in the clinic. Among our patients 3.1 per cent had this condition. The tumours were found in five men and two women whose ages ranged from 56 to 70 years. Since it is a disease which does occur and is of practical interest, we shall dwell here in detail on some of the cases.

Hypernephroma in our patients occupied the isthmus of the horseshoe kidney, or the right or left half of the kidney, or the right half and the isthmus. In two cases the tumour was found in a duplicated half of the fused organ, in one case, in addition, there was a retrocaval ureter. In half of the cases the renal tumour was attended with pyelonephritis.

Symptomatology. According to the clinical picture, two groups of patients with hypernephroma can be distinguished: patients with no symptoms of the disease, and those who have symptoms.

In one of our patients the renal disease followed an asymptomatic course.

S., a 70-year-old man, was admitted to the clinic on August 29, 1964, complaining of malaise, elevated body temperature, and constipation. In the last two months he had been treated for rheumatism. X-ray of the lungs performed at an out-patient clinic unexpectedly showed numerous metastases of a tumour. To determine its primary localization the patient was sent to the clinic. He was emaciated and the sclerae and skin had a yellowish tinge. The left cardiac border was shifted to the left, the cardiac sound diminished; a systolic murmur was heard at the cardiac apex and an accentuated second sound in the pulmonary artery. AP was 140/70 mm Hg. Percussion produced a bandbox sound over the lungs, auscultation revealed diminished breathing. The liver was enlarged. A hard, nodular, poorly mobile tumour was palpated in the right iliac region. Percussion over it elicited a dull sound. Congenital shortening of the left lower extremity was noted. Blood test: Hb 75 per cent; leucocytes 8700: eosinophils 1, rod neutrophils 1, filamented neutrophils 80, lymphocytes 10, monocytes 8; ESR 13 mm/hr. Urinalysis showed normal values. Chest X-ray confirmed the presence of many metastases of a tumour in the lungs. Excretory urography showed diminished functioning of both kidneys. Retrograde pyelography showed three branches arising from the right ureter at the level of the fifth lumbar vertebra, each with its own pelvis located above the fifth lumbar vertebra (Fig. 52).

This same ureter had still another pelvis at a higher level whose lower calyces had a filling defect. On the level of the fifth lumbar vertebra, to the right of the ureteral branching, another shadow of an irregular shape was seen, probably an accessory pelvis with a filling defect.

The following diagnosis was made: tumour of the right half of a horseshoe kidney with numerous metastases in the lungs and liver, duplication of the right half of the fused organ with splitting of the ureter. The patient died on November 15, 1964. The diagnosis was verified on autopsy. The tumour occupied (Fig. 53) the right half and the isthmus of the fused organ. Histological examination showed carcinoma of the kidney.

Fig. 52. Retrograde pyelogram. Hypernephroma of horseshoe kidney

In this case the tumour remained asymptomatic for a lengthy period of time, till signs of its metastases appeared in the lungs and liver. There was no pain or haematuria, and the tumour was palpated at a site uncommon for the kidney, i.e. in the right iliac region. Such an atypical picture of the disease, which was

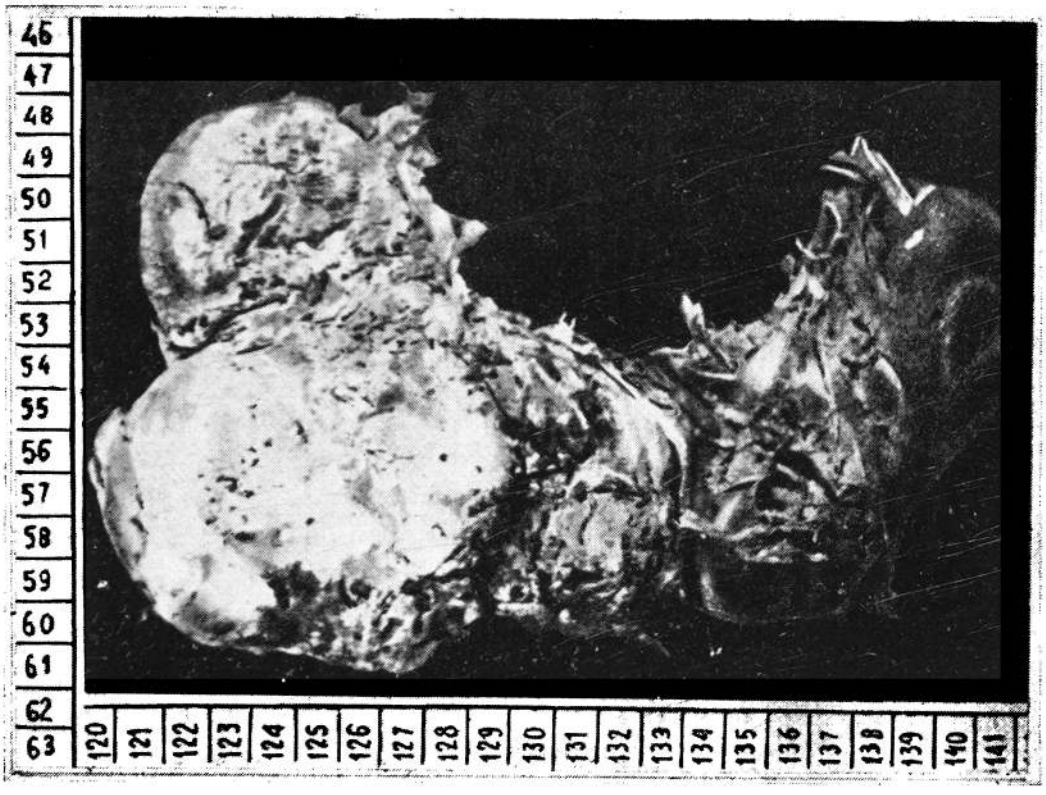


Fig. 53. Postmortem preparation of horseshoe kidney with hypernephroma in the right half and in the isthmus

also poor in symptoms, created considerable diagnostic difficulties. In one of their cases of nephrolithiasis Rose and Wattenberg (1945) unexpectedly found Wilms' tumour during histological examination of the removed isthmus of a horseshoe kidney. The manifestations of other diseases may sometimes screen the symptoms of the renal tumour. The symptoms of acute glomerulonephritis predominated in a case in Smith's practice (1947), for instance, whereas autopsy revealed that the patient also had a tumour of the kidney.

Various local symptoms of the renal tumour are found in some patients. In our practice, the most common symptom was a hard, nodular, and immobile tumour palpated in the abdomen on the midline.

Pain was localized at the umbilicus or in the iliac region. In half of the cases there was total haematuria. The generally known triad of symptoms of a tumour in a fused kidney was encountered in only one of our cases and even then it was marked by some peculiarities.

M., a 65-year-old man, was admitted to the clinic on April 8, 1958. He complained of permanent dull pain in the right iliac region and at the umbi-

licus and bloody urine. A hard, nodular and immobile tumour was palpated in the right iliac region. The percussion sound over it was diminished. Pasternatskii's sign was positive on the right. Blood test: Hb 78 per cent; leucocytes 5300: eosinophils 3, rod neutrophils 7, filamented neutrophils 65, lymphocytes 21, monocytes 4; ESR 55 mm/hr. Urinalysis: reaction acid, specific gravity 1020, protein 0.066 pro mille, leucocytes 5-7 in field of vision, erythrocytes 45-50 in field of vision. A pyelogram showed a low position of both pelves, which were located at the level of the fourth to fifth lumbar vertebrae and the sacroiliac articulation. Their calyces were stretched out and directed medially, those of the right pelvis had filling defects. A diagnosis of tumour of the right half of a horseshoe kidney was made. Histological examination revealed hypernephroma.

In this case pain was felt in the iliac region and at the umbilicus and the tumour was also palpated there. Haematuria was the only definite sign of a renal disease.

Other symptoms were rarely encountered. These included positive Pasternatskii's sign, pyuria, proteinuria, arterial hypertension, fever, cardiovascular disorders, changes in differential blood count.

Carcinoma of the kidney is marked by a chronic **clinical course.** Though the symptoms of the tumour had been manifested for a short period, the new growth itself was found to be large with metastatic spread to remote organs. This is evidently associated with the latent course of the disease, but it may be that carcinoma in a fused kidney has a particular tendency to produce metastases early and in a great number.

Diagnosis. A tumour palpated close to the vertebrae often provides grounds for suspecting a fused kidney. A changed percussion sound over the tumour should be disregarded because it may be diminished or tympanic. X-ray examination is the main diagnostic method.

A **scout urogram** sometimes shows an image of the fused organ and, in some cases, calcification of the tumour.

Excretory urography rarely gives a clear idea of the structure of the kidneys and urinary tract in elderly patients because of low renal function, the more so since the pelves of malformed kidneys are often concealed on radiographs by the shadows of the bones.

Retrograde pyelography yields more convincing data in the diagnosis of the anomaly and the disease in such cases. On the pyelograms we saw low position of the pelves, with the calyces facing the vertebrae and the short ureters. In this disease the X-ray signs of fused kidneys are attended with defects in the pelvis and calyces. The uncommon shape of the pelvis and calyces, however, creates difficulties in the diagnosis of the tumour of the kidney. In this case diagnostic **pneumoretroperitoneum** is helpful. The image of the fused organ is usually demonstrated clearly on the pneumograms.

The uncommon shape of the calyces of a malformed kidney is sometimes mistaken for signs of a tumour. There are also cases in which a tumour of other organs is recognized erroneously as tumour of the kidney. Taylor et al. (1953), for instance, mistook neuroblastoma of the lumbar region for a tumour of a horseshoe kidney. It is particularly difficult to differentiate carcinoma and polycystic disease of the kidney. Lateral view X-ray examination is very important in resolving such diagnostic doubts. This type of examination helped Perrier (1936) to detect a filling defect in the calyx which could not be seen on a posteroanterior radiograph. Excretory urography combined with tomography may also produce the necessary information. By combining these two methods of examination Dufour (1959) was able to make the diagnosis of tumour and a polycystic horseshoe kidney. Tumour of the kidney may be properly diagnosed on the grounds of the developed vascularization of a renal area as demonstrated by angiography. This examination is now preferred to the other methods in cases of tumour of fused kidney. Timely use of modern diagnostic methods makes it possible to recognize tumour of a fused kidney in all cases.

Solitary Renal Cyst

We dwell here on the combination of a cyst and a horseshoe kidney since it is encountered not altogether rarely. Among our patients there were three with a solitary cyst. In one of them it produced clinical symptoms and because of that it was of practical importance. The relative rarity of the condition and some peculiarities of the clinical picture and diagnosis warrant giving the case in illustration.

M., a 56-year-old man, was admitted to the clinic on March 4, 1967 complaining of blood in the urine. Haematuria had occurred several times within a month but the reason for it could not be found. An elastic, painless, and slightly mobile tumour was palpated below the umbilicus, more to the right. It was situated across the spine. Percussion elicited a tympanic sound over it. Blood test showed normal values. Urinalysis: reaction acid, specific gravity 1019, protein 1.65 pro mille, leucocytes 5-6 in field of vision, erythrocytes covering the field of vision. Indigo carmine was discharged from the left ureteral orifice in the fourth minute and from the right in the seventh minute. The image of a horseshoe kidney was seen on a scout radiograph. An urogram demonstrated both pelves on the level of the second to third lumbar vertebrae, the right one over the transverse vertebral processes and the left deviated from the vertebrae but with its calyces directed medially. Right retrograde pyelography showed the right pelvis above the third lumbar vertebra and its ureter displaced to the midline. The structure which seemed to be a pelvis on excretory urography was actually its upper calyx. The pelvis and lower calyx had a filling defect. The favourable course of the disease suggested a solitary cyst of a horseshoe kidney. The diagnosis was

verified on operation. A large solitary serous cyst was found in the lower pole of the right half of a horseshoe kidney. The cyst was excised. Histological examination showed that its wall was of fibrous tissue.

In this case painless profuse haematuria disclosed the disease. An uncommon localization of the tumour at the umbilicus helped to suspect a horseshoe kidney, while the favourable course of the disease was suggestive of a solitary cyst. The findings of excretory urography and retrograde pyelography confirmed the diagnosis.

TREATMENT OF FUSED KIDNEYS AND THEIR DISEASES

Non-operative Treatment

The purpose of the non-operative treatment of fused kidneys is to relieve pain and neurological disorders which may develop as a consequence of compression of the nerve plexuses and pathological mobility of a fused organ that is not complicated by a disease. Mild manifestations of the anomaly may be relieved by prescribing bed-rest for a few days, analgesics, baths, and a heat pad. More intense pain is alleviated by a procaine (0.5 per cent solution) blockade in the region of the spermatic cord or the round ligament of the uterus. When there is stasis of urine in the upper urinary tract due to compression of the ureter above the isthmus of a horseshoe kidney, temporary relief may be gained by catheterization of the pelvis. The following case illustrates this type of treatment.

P., a 49-year-old woman, was admitted to the clinic on October 4, 1966. She complained of dull pain in the epigastrium and right hypochondrium experienced from time to time over the last three years. On examination an elastic, painless, and hardly mobile tumour was palpated in the region of the umbilicus. Blood and urine tests showed normal values. The image of a horseshoe kidney was seen on a scout radiograph. A pyelogram demonstrated both pelves at the level of the third to fourth lumbar vertebrae; they were surrounded by their calyces, which were included in the 'lumbar rectangle'. Diagnostic pneumoretroperitoneum showed a clear image of a horseshoe kidney. No disease in the anomalous kidney was found. Bed-rest, Analgin (Dipyrone), and a heater were prescribed. After such measures and, in addition, catheterization of the right pelvis all the symptoms abated. On discharge from the clinic the patient was recommended to restrict muscular exertion. During a three-year follow-up the episodes of pain recurred twice.

We had the opportunity to study the results of non-operative treatment of 23 patients with fused kidneys not complicated by disease. In most cases they proved satisfactory. The morbid manifestations were rare and dull and sometimes sharper pain troubled the patients only for short periods. There were no func-

tional or morphological abnormalities in the kidneys, and patients never needed to miss more than a few days' work.

In some cases the pain became more intense each time and occurred more frequently, as the result of which the sick person had to leave work more often, although the anatomical and functional condition of the fused kidneys was within normal values. Non-operative treatment proved ineffective in such cases.

Non-operative treatment is sometimes also applied in the initial stages of hydronephrosis and in nephrolithiasis, pyelonephritis, tuberculosis and nephroptosis.

Catheterization of the pelvis in the initial stages of hydronephrosis has a favourable effect. The patient has no complaints, pyuria ceases, the normal volume of the pelves and calyces is restored, good renal function persists, and the patient can resume work. Such favourable results, however, are temporary; the disease will recur in two or three months.

In some cases of non-destructive forms of tuberculosis of fused kidneys non-operative treatment may be very effective. The following case from our practice is given in illustration.

Sh., a 57-year-old man, was admitted to the clinic on February 2, 1967. He complained of dull pain in the left lumbar region, frequent and painful micturition, and elevated body temperature. A diagnosis of pulmonary tuberculosis had been made in 1955 and of tuberculous epididymitis on the right side in 1962. He had undergone epididymectomy in 1965. Lumbar pain and signs of dysuria developed in 1966. On examination an elastic, painless and poorly mobile tumour was palpated to the left of the umbilicus. Blood test: Hb 78 per cent; leucocytes 8200: eosinophils 2, rod neutrophils 3, filamented neutrophils 76, lymphocytes 15, monocytes 4; ESR 15 mm/hr. Urinalysis: reaction acid, specific gravity 1015, protein 0.099 pro mille, leucocytes 100-200 in field of vision, erythrocytes up to 10 in field of vision. *Mycobacterium tuberculosis* were found in the urine by the floatation method. Indigo carmine was discharged from the right ureteral orifice in the fifth minute and from the left orifice in the tenth minute. The image of a horseshoe kidney was found on a scout radiograph. Excretory urography showed, against the background of a horseshoe kidney, a renal pelvis on a level with the second lumbar vertebra with the calyces pointing away from both sides of the ureter; wide spherical shadows of many calyces were visible to the left of the second and third lumbar vertebrae. A diagnosis of tuberculo-infiltrative tuberculosis of the left half of a horseshoe kidney was made. *Mycobacterium tuberculosis* were discharged in the urine. The patient received 37 g of cycloserine, 29 g of Saluzide (Opiniazide, carboxyverazid), 21 g of Larusan (isonicotinic acid), and 90 g of Ethoxyde (Etocarlide). On examination two years after the beginning of treatment the patient had no complaints and blood and urine tests showed normal values. Indigo carmine was discharged from both ureteral orifices in the fifth minute. Both pelves were clearly seen on a urogram. The left pelvis had reduced to normal size, it was small and elongated and its calyces were not dilated.

It can be seen from this case that non-operative treatment may produce good results in tuberculo-infiltrative tuberculosis of a horseshoe kidney. That is why we cannot agree with Pilkh (1968)

who contends that the efficacy of antituberculous treatment is considerably reduced in affection of a horseshoe kidney.

Because of chronic stasis of urine in the pelves of fused kidneys, non-operative treatment of nephrolithiasis, pyelonephritis and nephroptosis has satisfactory results only at the time when it is applied. For this reason we consider treatment of patients with nephrolithiasis of fused kidneys at a spa inadvisable, since balneotherapy in cases of chronic stasis of urine in the pelves is conducive to the progression of the disease.

We encountered in our practice several patients with nephrolithiasis attended with considerable stasis of urine and infection, as well as patients with renal tuberculosis and attendant nephrolithiasis and hydronephrosis, who could not agree to surgery. In these cases the results of non-operative treatment were poor.

Non-operative treatment does not arrest the advance of hydronephrosis and pyelonephritis. Three of our patients with bilateral hydronephrosis and pyelonephritis died from renal insufficiency. They had been brought to the clinic in an extremely grave condition.

Surgical Treatment

Indications and Contraindications

In morbid disorders of fused kidneys some authors apply surgery (Foley, 1940), while others consider it inexpedient because the outcomes of the operations do not justify the surgical risk (Culp and Winterringer, 1955). We believe that an operation on a fused kidney not complicated by disease is indicated when the patient has permanent dull pain or frequent attacks of sharp pain which make him incapable of working and which are usually attended by considerable motor disorders of the upper urinary tract. These symptoms evidently indicate the onset of a disease. Non-operative treatment in such cases is usually ineffective.

According to many surgeons, the indications for the surgical management of diseases of fused kidneys do not differ from those applied in cases of the affection of normal kidneys with these diseases (Bono et al., 1962; Nelzina, 1962). Some surgeons believe that chronic stasis of urine in the pelves of fused kidneys is conducive to poor outcomes of operative interventions. That is why they recommend restriction of the indications for surgery (Lyakhovitsky and Sukhanov, 1936; Smirnov, 1959). Others think that surgery is advisable when non-operative treatment is ineffective and there is urinary stasis in the pelves (Mombaerts, 1961).

It is our experience that two types of indications should be considered, those in diseases characterized by urinary stasis, and those in diseases without urinary stasis. In the latter case they do not differ from the commonly accepted indications. In considering the indications for surgery it is necessary to take into account the severity and character of the clinical symptoms and the functional and morphological changes in the kidney, the presence and virulence of infection, the character and localization of stones, the causes of the disease, and the patient's general condition. The high frequency of mechanical causes of the development of diseases of fused kidneys should be borne in mind, as well as the frequent occurrence of bilateral affection and the combination of several diseases in one half of the kidney and the fact that in unilateral affection it is highly probable that the disease may ultimately develop on the contralateral side. It is for these reasons that we believe that indications for operations on fused kidneys, particularly in bilateral affection, should be considered on a wider scale, particularly as non-operative treatment is ineffective in such cases and the disease tends to develop further.

In considering the indications for surgery in such cases it should also be borne in mind that the tumour or inflammation may spread from one half of a fused kidney to the other by way of the isthmus. We encountered this in cases of tuberculosis and tumours.

Patients with hydronephrosis should be treated by surgery. We found it necessary to operate on such patients when they experienced permanent dull pain or attacks of sharp pain attended by periods of pyelonephritis which made them incapable of working, as well as when the disease progressed and was marked by severe hydronephrosis with disturbed renal function, and particularly when the affection was already bilateral.

We found surgery to be expedient when nephrolithiasis was manifested by continuous dull pain or frequent attacks of renal colic, as a result of which the patient could not work, as well as in calculous pyelonephritis, hydronephrosis and pyonephrosis. We believed it wrong to wait for long-term spontaneous discharge of the concrement, because this rarely occurs in cases of stones in a fused kidney. In elderly patients with aseptic stones and no marked clinical manifestations of the disease, the indications for surgical management are of a relative character, if the stones do not cause disorders of renal function. Temporizing tactics is admissible in such cases.

With recurrent pyelonephritis only an operation can relieve urinary stasis and prevent further development of the disease, which is why pyelonephritis of a fused kidney with severe sta-

sis of urine in the pelves should be regarded as an indication for surgery. A wider range of indications for surgical treatment should be considered in cases of combined diseases.

There are no special contraindications for surgery of fused kidneys. It should only be noted that the immobility of the tumour should not be accepted as a criterion of its inoperable nature because fused kidneys are usually poorly mobile.

Special Operative Techniques in Manipulations on Fused Kidneys

Various operations càn be performed on fused kidneys. According to technique, the following two groups can be distinguished: operations without transection of the renal isthmus and operations with transection or resection of the isthmus of the anomalous organ. The operative interventions in the first group differ little from those commonly employed in the management of renal diseases. Operations of the second group are performed only in cases of this renal anomaly.

Approach to the kidney. It is sometimes very difficult to operate on a fused kidney because of its deep localization, poor mobility, attachment to the spine by the isthmus and accessory vessels, and the variability of its anatomical structure. These difficulties can be avoided to a considerable extent by using the right approach to the kidney. Good anaesthesia is very important in such cases. Intubation anaesthesia with the use of muscle relaxants is recommended.

Several variants of approach to a fused kidney are described in the literature, but all have their own shortcomings. Some surgeons operate on a fused kidney by means of an intraperitoneal approach (Bazhenov, 1961; Parker, 1965), others prefer the extraperitoneal lumbar (Gregoir, 1963; Shatin, 1963) or a combined (Kudryavtsev, 1967) approach. The expediency of using various methods of access is still the subject of discussion.

Each method of approach to the kidney should meet several requirements. It should be simple, universal and should allow close inspection of the kidney and its isthmus. We find that the extraperitoneal approach meets these requirements best.

The extraperitoneal lumbar approach is made as follows. The patient is placed on his healthy side with the trunk drawn back to an angle of 45 degrees. The lower limb on the healthy side is drawn up at a straight angle, the other limb is stretched out and a bolster is placed between them.

An oblique transverse incision is made. For better inspection of the whole isthmus and part of the contralateral half of the

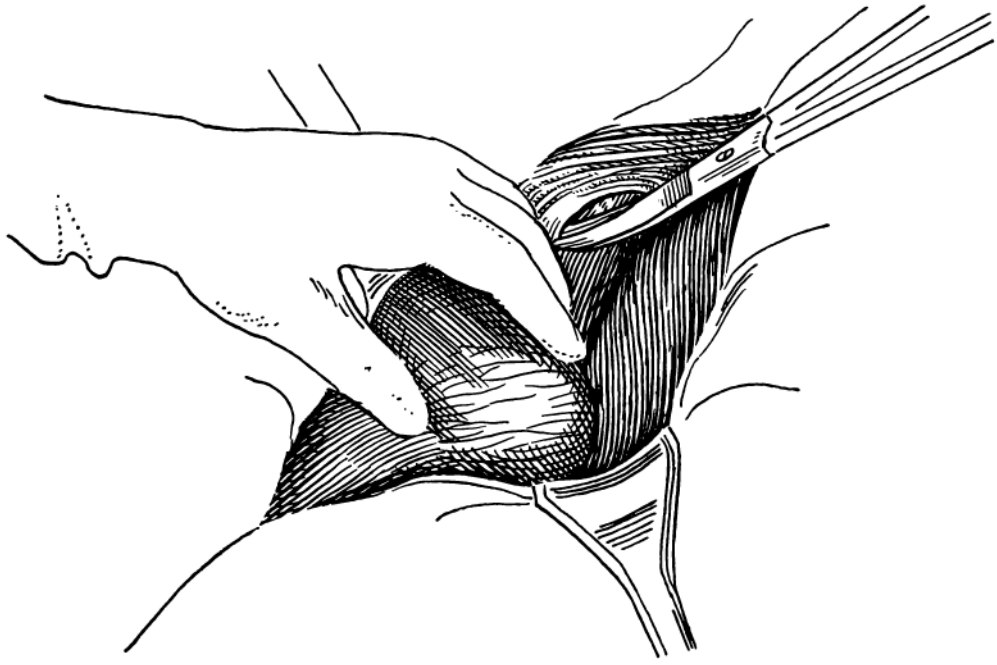


Fig. 54. The peritoneum and perirenal fascia are moved to the front as far as possible and the renal fascia is cut

kidney it is sometimes necessary to partly transect the sheath of the rectus abdominis muscle.

Since the peritoneum covers the anterior and lateral surfaces of the anomalous kidney, it has to be moved to the midline as far as possible and the retrorenal fascia cut more to the back at the lumbar muscles (Fig. 54).

It is simpler and easier to begin exposure of the fused kidney at its smooth posterior surface, which is usually freed from the surrounding tissue without any difficulty. As a rule the posterior surface of such a kidney has no gross adhesions with the adjacent organs. However, small accessory vessels draining directly into the renal substance may be found here, which is why the posterior surface of the kidney should be exposed carefully under visual control.

When the posterior surface of the kidney has been freed to its medial margin, a finger is passed around it and its tip is then palpated through the perirenal fat medially of the pelvis (Fig. 55). The peritoneum, fascia, and perirenal fatty tissue are removed from the anterior surface of the kidney, with the finger as a guide. The finger holds the kidney in the wound and pulls it away from the large abdominal vessels. It is very difficult to hold it for very long in this manner, since the hand tires easily and hides the field of vision. So we use for this purpose a special

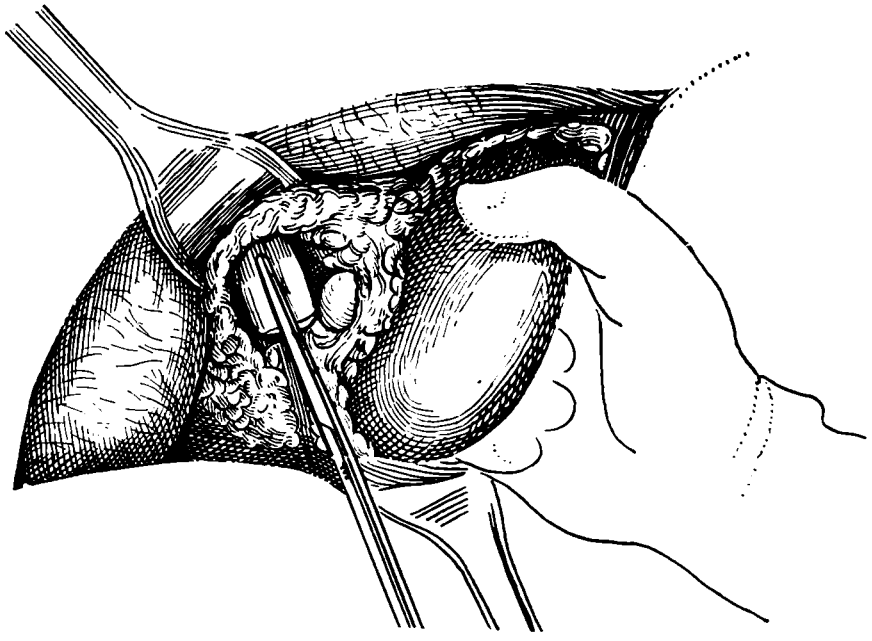


Fig. 55. The posterior even surface of the horseshoe kidney is first freed and then the kidney is pulled out into the wound with the hand. The fatty tissue and peritoneum are removed from it using the finger as a guide

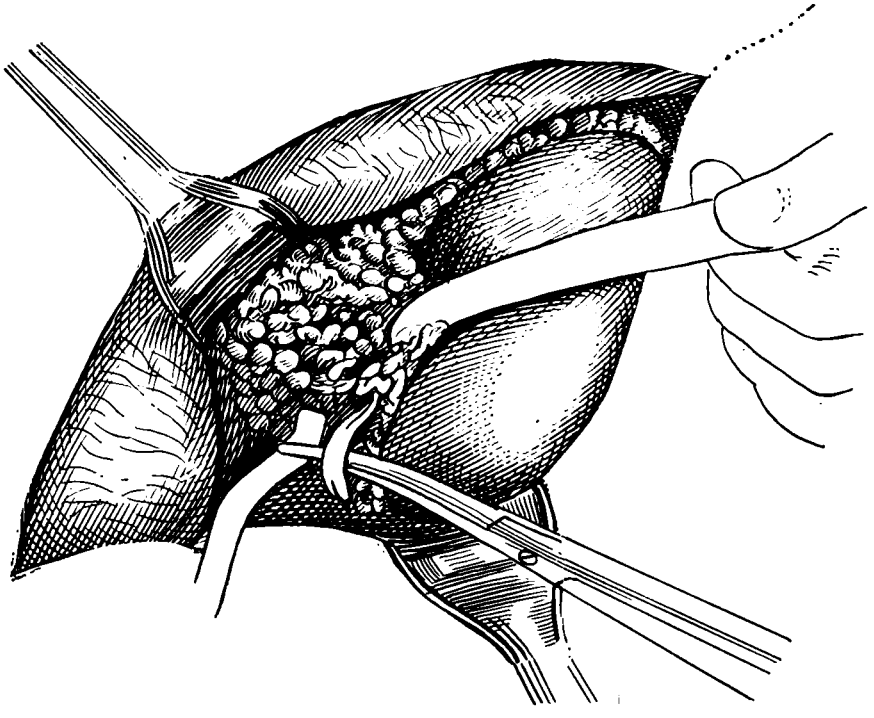


Fig. 56. The horseshoe kidney is drawn out into the wound with a gauze loop. The second loop is passed about the ureter

gauze loop passed around the kidney along the finger (Fig. 56). It draws the kidney out into the wound well and makes further exposure of the isthmus easier.

When there is no need to expose the isthmus of a horseshoe kidney, for instance in routine pyelolithotomy, the necessary surgical manipulations are begun as soon as the anterior surface of the kidney is freed.

In a reconstructive operation the ureter is separated from the surrounding tissues at the lower pole of the kidney and held with an additional loop. Further exposure of the isthmus is often prevented by accessory vessels which reach the lower pole and the isthmus from the aorta and iliac arteries. They are usually of little importance in the vascularization of the kidney, but sometimes such a vessel may supply blood to a large area of the lateral part of the kidney. Zones supplied by these vessels are determined by means of simple tests. The vessel is compressed and the area with disturbed circulation is revealed by its dark colour. In cases of well developed collateral vessels in the renal isthmus, however, this test is ineffective. In such a case one compresses the vessel and the isthmus at the same time (Fig. 57), as a result of which

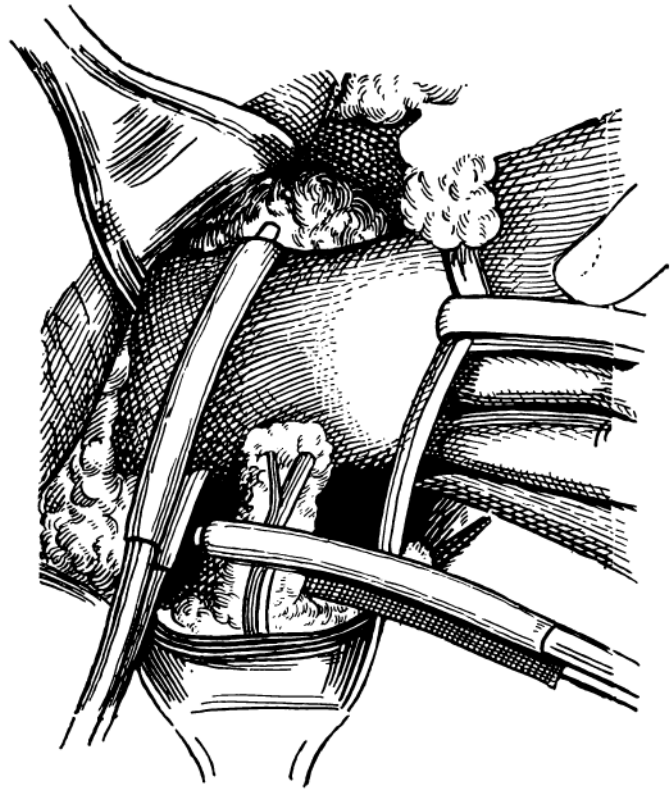


Fig. 57. Clamps are applied to the accessory vessels at the lower pole of the kidney and to the isthmus

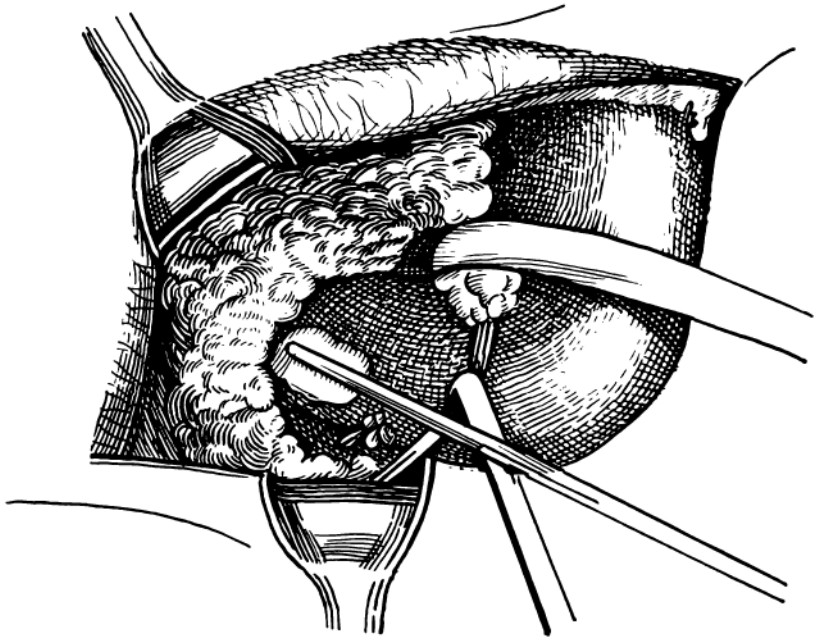


Fig. 58. The accessory vessels are transected between two clamps. The vessels at the lower pole of the kidney are ligated.The isthmus is exposed

the zone with impaired circulation is clearly seen. This manipulation helps the surgeon to avoid an error and to preserve a vessel that is important in supplying blood to the kidney.

After transection and ligation of the accessory vessels the isthmus is easily separated from the surrounding tissue on all sides (Fig. 58) and an additional gauze loop can be passed around it, with which it is drawn out well into the wound. These manipulations not only provide a good approach to the isthmus of a horseshoe kidney, but make it possible to examine part of the contralateral half of the anomalous kidney through the oblique lumbar incision.

In asymmetrical types of fused kidneys the extraperitoneal approach is still easier and resembles the approach to a normal kidney.

We prefer the extraperitoneal approach in various operations, including reconstructive operations with transection or resection of the isthmus and part of the lower pole of a horseshoe kidney. A skilled surgeon does not need much time to gain access to the kidney by this method and if necessary may proceed from simple manipulations to more complicated ones.

The intraperitoneal approach is usually made by means of median laparotomy through an avascular area of the mesentery or through the peritoneum lateral to the descending colon. In the latter case the intestine is moved to the midline together with

the tail of the pancreas. Transverse incisions are also used or a thoracoabdominal incision is made between the tenth and eleventh ribs on the left or the eleventh and twelfth ribs on the right and continued in the transverse direction toward the umbilicus. If necessary, a combined incision is made.

We have used this intraperitoneal approach for removing a localized tumour of the isthmus of a horseshoe kidney. The parietal peritoneum was cut in the mesenteric area poorest in vessels. We nevertheless prefer the extraperitoneal method because we feel that with definite skill it does not take much time, enables one to proceed from simpler to more complicated manipulations whenever necessary, and is less dangerous as regards infection of the abdominal cavity.

Operations without transection of the isthmus of a horseshoe kidney. Simple operations without transection of the isthmus of a fused kidney hardly differ in technique from operations performed on normal kidneys. The poor mobility of the fused kidney has to be taken into account and the operation is carried out without drawing the kidney out into the wound. Since the pelves are usually located on the anterior surface of the organ, anterior pyelolithotomy is performed for nephrolithiasis. It is best to make the incision in the lower part of the renal pelvis, which in most cases is devoid of vessels. During the operation it is necessary to examine the calyx which is directed medially and at the isthmus, because a stone may be left there.

It is particularly difficult to reoperate on a fused kidney. The poorly mobile and deeply located kidney is joined by adhesions to the surrounding tissues and has to be cut out of the gross scars with scissors. The peritoneum covering the entire anterior surface of the kidney and the numerous unusually located vessels may be damaged during this manipulation. It is advisable to expose the pelvis directly at the upper margin of the isthmus where there are no vessels.

It is our opinion that in nephropexy high anchoring of the kidney should not be attempted, so as not to stretch the short anomalous vessels. To restrict the mobility of such a kidney it is sufficient to attach it by the capsule to the lumbar muscles by means of a few interrupted sutures.

Operation with transection of the isthmus. Operation on a fused kidney with transection and especially with removal of its isthmus is much more difficult than operations on a normal kidney. The difficulties arise because the kidney cannot be drawn out into the wound and because the topographic-anatomical localization of the organ and its urinary tract and vessels is abnormal, the renal pedicle is short, there are numerous vessels, and the aorta and vena cava are found in close vicinity. Moreover,

the technique of such operation is as yet insufficiently developed.

The **choice of the site for the transection of the kidney** or of the zone for its resection determines the results of the operation to a great extent. It is a simple matter to transect a fibrous isthmus. But an isthmus of fibrous tissues is a rare occurrence. In our practice we encountered only parenchymal isthmi of horseshoe kidneys, which were 1 to 6 cm in length and breadth and 1 to 2.5 cm in thickness. Under such conditions the site of the transection has to be chosen within the boundaries of the tissue, at the narrowest part or at the lower pole of the kidney at some distance from the renal pelvis. Quite often it becomes necessary to resect the calyx together with the isthmus during a reconstructive intervention. In several of our cases this was not followed by the development of a fistula. This complication is evidently due to urinary stasis persisting in the pelvis after the operation rather than to any deficiency in the sutures applied to the transected calyx.

Much importance is attached to the structure of the vascular network within the anomalous organ we have studied at autopsy. On analysing the results of our examinations we have tried to find parallels between the structures of the extrarenal and intrarenal arterial networks, to correlate the intrarenal distribution of the vessels with the shape of the kidney and with the segmental sulci often found on its surface. We encountered lobate kidneys in twelve autopsies.

We studied intrarenal arterial circulation in eight kidneys, the vessels of which were shown on radiographs. In three cases the number of arteries was in accordance with the number of vascular segments; in the other cases there were more segments than vessels and they coincided with the lobate structure of the kidney. In two cases the isthmus of the horseshoe kidney had good anastomoses between the vessels of the lateral renal lobes; in three cases there was a relatively avascular zone at the fusion of the kidneys; and in still another three the isthmus contained a vascular segment, which usually occupied not only the isthmus but the nearest part of the lower pole of the kidney as well. Thus, the accessory vessel of the isthmus or lower pole provides a small area of the kidney here with blood. Sometimes, however, such an accessory vessel effected the blood supply to a large part of the fused organ. In one of our cases accessory vessels created large segments in the lower poles and the lateral parts of the kidney. In this case on an angiogram vascular anastomoses were clearly seen in the isthmus of the horseshoe kidney.

Experience in our practice suggests that in most horseshoe kidneys the principle of segmental distribution of the intrarenal arteries is preserved despite the great variety in the external

structure and the vascular bed in this renal anomaly. The segments mentioned correspond to the lobate structure of the kidney. The more conspicuous the lobate structure of the kidney, the more clearly are the boundaries of the vascular segments defined. This feature confirms the validity of economical operations on horseshoe kidneys and can be put to use in the clinic.

Judging from the available data, it is most advisable to transect a horseshoe kidney along the sulcus found between the lobes which forms the boundary of the vascular segments. When the isthmus is not supplied with accessory arteries, one should expect there to be no large vessels in its middle portion and that its division or excision will not disturb circulation in the poles of the kidney. When the isthmus or lower pole has accessory vessels, a vascular segment is usually located here with its boundaries extending to the lower pole. In such cases, after ligation of the accessory vessel excision of the isthmus with part of the lower pole is advisable, so as not to leave large renal areas with disturbed circulation. Sometimes the accessory vessels supply blood to a large part of the kidney. It is therefore necessary to determine the zone of vascularization before ligating these vessels. Vascular segments present in the lateral lobes may be resected without any great risk of causing circulatory disorders throughout the whole kidney.

Methods of transecting or resecting a fused kidney also affect postoperative outcomes. Transection or resection of the isthmus of a fused kidney is the most crucial stage of heminephrectomy and reconstructive manipulations and is the cause of most of the severe complications, such as haemorrhage and urinary fistulas.

Most surgeons divide the kidney between two rows of catgut mattress sutures (Lowsley, 1952); others compress it beforehand with soft clamps (Shatin, 1963) or crush it along the site of the transection with an écraseur, then apply two clamps to the thinned out area, dividing the kidney between them, and then place a haemostatic suture at the sites of the incisions (Baker and Colston, 1963). This, nevertheless, proves insufficient for arresting bleeding, so the renal wound has to be covered with the peritoneum, omentum, fatty tissue, muscle or plates of polyvinyl alcohol or oxidized cellulose (Elansky, 1939; Davidson, 1941; Gekhman, 1962), and the kidney divided with an electric knife (Mikhnovskaya, 1959).

We have had the opportunity to study different methods of resection of the isthmus of a horseshoe kidney. In our first ten cases we divided or resected the isthmus by the following commonly accepted methods: (1) between two rows of catgut mattress sutures; (2) between two rows of catgut mattress sutures with preliminary application of clamps to the kidney; (3) routine wedge

resection. In heminephrectomy application of a single row of haemostatic sutures and a clamp to the part which will be resected proves sufficient. In a reconstructive organ-preserving operation two rows of haemostatic sutures have to be applied at a distance of 2.0 to 2.5 cm one from each other, and the kidney divided between them.

To prevent the haemostatic sutures from slipping off the renal parenchyma the kidney is cut at some distance from them. As the result of this a large renal area with disturbed circulation remains in the wound, which clearly affects healing of the wound and the course of the postoperative period. In such cases bits of fatty tissue or 'broken' muscle are inserted under the sutures, which prevents them from cutting through and improves haemostasis. In addition, stitches should be placed around some of the vessels in the renal wound and tied. All this complicates the operation and takes much time.

In eight of these ten operations good haemostasis was produced. A small retroperitoneal haematoma developed in one case (there was a sanguineous discharge from the wound for three weeks), and profuse haematuria occurred for three days in another. Although reoperation was not necessary in either of these cases, the complications aggaravated the postoperative period considerably.

The shortcomings of technique in these operations and the inadequacy of haemostasis in some cases prompted a search for other methods of dividing the isthmus of a horseshoe kidney. For this purpose we studied the possibility of using a mechanical tantalum suture applied by means of the lung root stitcher (УКЛ-60).

There is no information in the literature on the morphological changes in the kidney after the placement of the mechanical tantalum suture, the fate of the staples in the renal parenchyma or the healing period as compared to that in the commonly accepted technique of wound closure. We studied the morphology of renal wound healing after resection of the kidney by means of the mechanical stitcher, an electric knife, and after routine wedge resection in experiments on 25 animals. It was established that in terms of healing operations in which the mechanical tantalum suture was used did not differ from wedge resection of the kidney and excelled resection made with an electric knife. After application of the mechanical tantalum suture all morphological changes are restricted to the zone of the resection, the staples are fixed tightly on the renal capsule and do not promote chronic inflammation or the formation of stones, while the vascular network in the zone of the resection hardly differs from that of the contralateral kidney which had not been operated on.

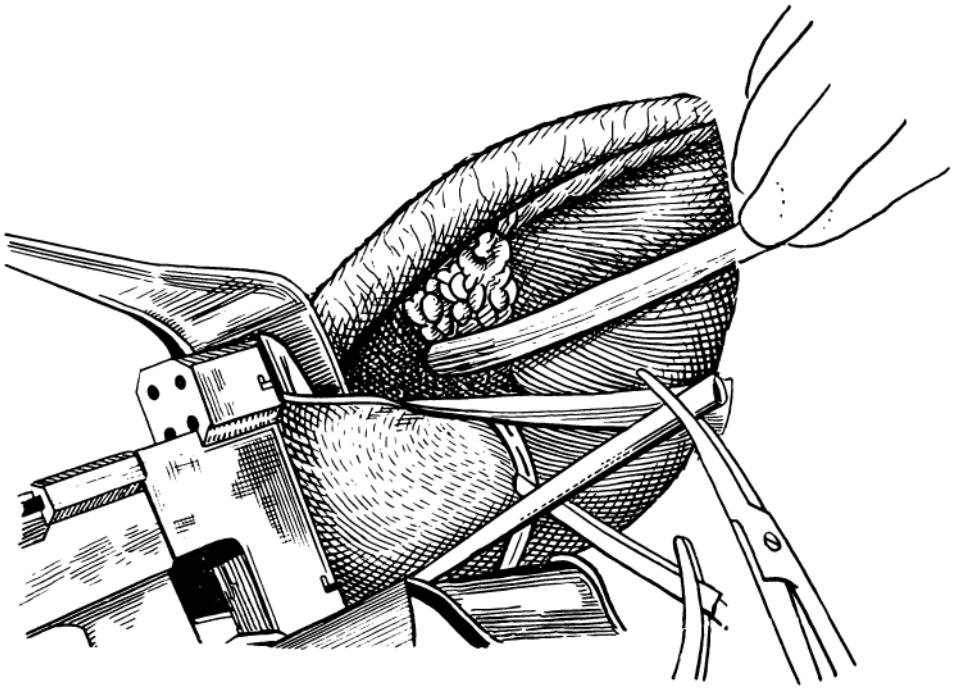


Fig. 59. The isthmus of the horseshoe kidney is drawn out into the wound with an additional loop. The blades of the УКЛ-60 stitcher are brought together as tightly as possible and tantalum staples are stitched through the kidney

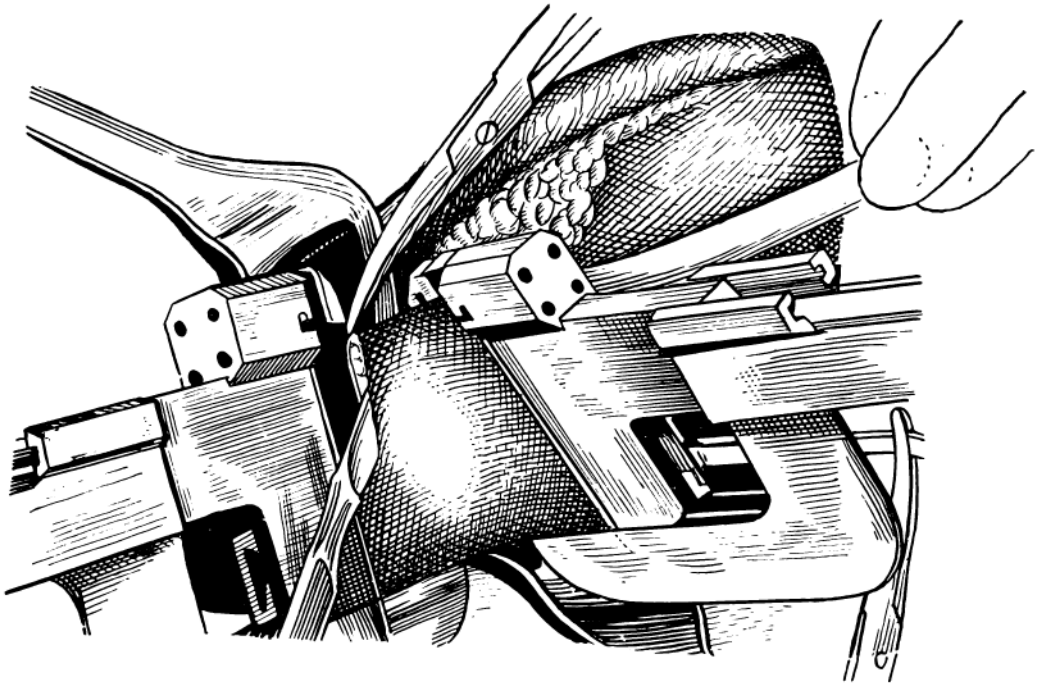


Fig. 60. Resection of the isthmus of the horseshoe kidney between two УКЛ-60 stitchers. The divided part of the kidney is grasped with forceps for its fixation in the wound

The results of the experimental research encouraged us to use the method in the clinic. We applied the mechanical tantalum suture in 46 operations on 41 patients. In all of them the horseshoe kidney was found to have a parenchymal isthmus. Histological examination revealed various changes characteristic of hydronephrosis or pyelonephritis in some isthmi and no abnormalities in others.

The operative technique with the mechanical tantalum suture is as follows. The renal isthmus is exposed with the capsule and, in a repeated operation, with some of the postoperative scars. They make the staple suture stronger. The isthmus, freed from the surrounding tissues, is placed into the open blades of the УКЛ-60 stitcher. During this manipulation the isthmus is raised and pulled away from the vessels with a loop. After that by rotating the screw to the right the isthmus is pressed between the blades as much as possible, leaving a clearance of 2.5 mm between the blades. The tantalum staples are then stitched through the kidney (Fig. 59). Both sides of the area to be removed are treated in this way. When two stitchers are used, they are fitted to both sides of the isthmus (Fig. 60). The kidney is then divided at some distance from the blades, so that the capsule can

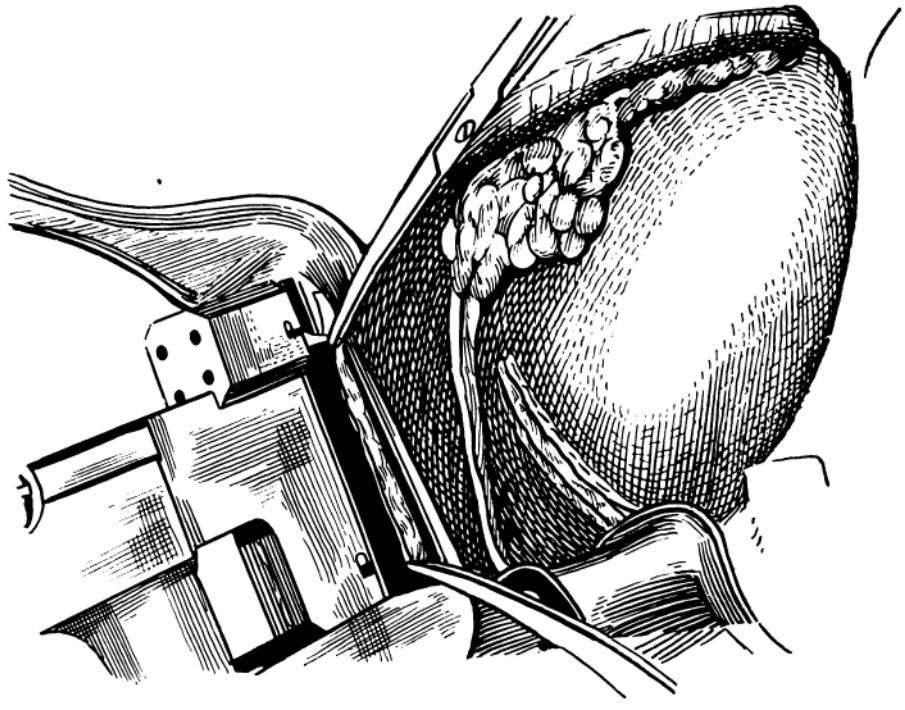


Fig. 61. The stitcher is removed from the lower pole of the left kidney. The capsule of the right kidney is held with forceps before removing the other stitcher

be grasped with two forceps, which hold the kidney in the wound while the stitcher is removed from it (Fig. 61). If necessary, this is followed by plastic procedures, pyelolithotomy or nephropexy on the renal pelvis. A drain is left in the lower pole of the kidney at the end of the operation or a tampon is sometimes put on the site of the resection for 24 hours.

The mechanical tantalum suture ensured reliable suturing of the renal tissue of the isthmus, good haemostasis and air-tightness of the calyces. In none of our cases did we encounter haemorrhage, retroperitoneal haematomas or urinary fistulas in the postoperative period, although we often divided a renal calyx in the isthmus by this method. All our patients recovered and are kept under observation (follow-up periods of up to six years). Surgery performed on the contralateral half of the kidney two to six months after the first operation showed that the lower pole of the kidney could be easily exposed to the level of the scar with the staples. The staples were tightly fastened on the renal capsule and did not penetrate the renal tissue. This suggests that the wound healed without the development of haematoma or purulent inflammatory reaction in the surrounding tissues.

The following case illustrates the use of the УКЛ-60 stitcher for dividing the isthmus of a horseshoe kidney.

B., a 52-year-old man, was admitted to the clinic on February 26, 1961. He complained of continuous dull lumbar pain over the last 15 years and frequent attacks of acute pain in both lumbar regions attended by vomiting, abdominal distention, fever, chills, and frequent and painful micturition. Small concrements were discharged at time during urination. In 1957 he underwent right pyelolithotomy. Pasternatskii's sign was positive. Blood test: Hb 84 per cent; leucocytes 5700: eosinophils 2, rod neutrophils 3, filamented neutrophils 69, lymphocytes 21, monocytes 5; ESR 36 mm/hr. Urinalysis: reactions alkaline, specific gravity 1012, protein 1.65 pro mille, leucocytes and erythrocytes covering the entire field of vision. Urine culture produced growth of staphylococcus. Urine collected from both pelves contained 50-100 leucocytes in field of vision. Indigo carmine was discharged from the left ureteral orifice in the eighth minute and from the right orifice it was not discharged at all after 12 minutes. A scout radiograph showed the shadow of a concrement on the projection of the left kidney and the shadows of three large concrements on the projection of the right kidney. Retrograde pyelography showed signs of a horseshoe kidney with both pelves enlarged. The following diagnosis was made: bilateral hydronephrosis, pyelonephritis, and nephrolithiasis of a horseshoe kidney. Left anterior pyelolithotomy was carried out on April 10, 1961. The stone was removed. The postoperative period was uneventful. The patient was discharged from the clinic but later readmitted because of pain of the same character as described above in the right abdomen. An operation was performed on the right side on October 24, 1963. An oblique lumbar incision was made on the right through which the parenchymal isthmus of a horseshoe kidney 4×2 and 5×1.5 cm in size was exposed. The УКЛ-60 stitcher was fitted to the isthmus. The isthmus was compressed, stitched through with tantalum staples, and divided. The right kidney

Fig. 62. Tantalum sutures have been applied to the lower pole of the right kidney and the stitcher has been removed (photo taken during operation)

became mobile and was easily shifted away from the midline. The stitcher was removed from the isthmus of the left kidney, being held in the wound during manipulation with forceps. The haemostasis was good. The left kidney was fitted under a peritoneal sac. The УКЛ-60 stitcher was also applied to the lower pole of the right kidney which was then compressed, stitched through, and divided (Fig. 62). The prepelvic portion of the ureter was freed from scars, while pyelolithotomy with removal of the stones was carried out. The wound in the pelvis was closed with interrupted catgut sutures. The postoperative period was uneventful.

We compared 46 operations in which the mechanical suture was used with 10 operations in which manual sutures were applied and found that the postoperative period was more uneventful in the first group. This is probably explained by less operative

trauma, since these operations took a much shorter period of time. A certain role may also be played by the fact that no large renal areas with disturbed circulation are left after the mechanical suture.

The advantages of the use of the mechanical tantalum suture in resection of the isthmus of a horseshoe kidney are obvious. It is simpler, considerably reduces the time needed for the operation, does not leave large renal areas with impaired circulation, and obviates the need for additional treatment of the divided calyx. The staples are clearly seen on scout radiographs so that the position of the kidneys can be studied in the postoperative period without resorting to contrast techniques.

At the same time some shortcomings of the method should be noted. The mechanical tantalum suture can only be used in the resection of a kidney not thicker than 2.0 to 2.5 cm. On compression of a thicker isthmus the capsule may be ruptured by the strained parenchyma (as occurred in some of our animal experiments). This reduces the strength of the staple suture. This restriction reduces the value of the mechanical suture in the resection of normal kidneys. Its use is most advantageous in the resection of the isthmus of a horseshoe kidney, because the isthmus in most of these cases is less than 2 cm thick. Since only kidneys that are no more than 6 cm in breadth can be fitted between the blades of the stitcher it cannot be used for dividing wider isthmi. These are, however, very rarely encountered.

Because this method is simple, reduces the time needed for the operation, and, besides, is reliable and causes no complications, it can be recommended for operation on horseshoe kidneys.

Heminephrectomy. It is usually more difficult to perform heminephrectomy on a fused kidney than on a normal kidney because in the first case the manipulations are carried out on a hardly mobile organ with many accessory vessels, and also because it is necessary to divide the renal isthmus.

It is advisable to divide the isthmus of the horseshoe kidney first and then ligate the vessels and the ureter. We find that this makes the operation easier. To prevent complications in cases of tumours and tuberculosis, it is best to resect, together with the half of the horseshoe kidney, the whole isthmus to the level of the lower pole of the contralateral half. In cases of marked hydronephrosis with changes in the corresponding half of the isthmus, the half of the kidney is resected together with the involved half of the isthmus. But if no changes are found in the isthmus, the kidney is divided through the lower pole of the half that is to be removed.

Reconstructive organ-preserving operations. This group of op-

erations includes division or excision of the isthmus of the horseshoe kidney, sometimes with part or the whole lower pole of the organ, pyelolithotomy, and plastic procedures on the upper urinary tract. These manipulations are carried out in various combinations. The surgeon who undertakes these complex operations must be acquainted not only with the main anatomical features but also with the diverse variants of the anomaly, so as to be aware of all the unusually complicated relations.

It is best to begin the operation by dividing the fused organ into two separate kidneys. In resection of the isthmus the accessory vessels at the lower pole of the kidney and the vessels of the isthmus are ligated; the kidney is then divided or the isthmus excised, or the isthmus is resected together with part of or the whole lower pole of the kidney. When necessary, a plastic operation may then be undertaken for correcting the obstructed drainage of urine from the renal pelvis. During the plastic procedures the pelvis can be examined and stones removed.

To provide the upper urinary tract with a position that is more favourable for its functioning, the prepelvic part of the ureter should be relieved of compression above the isthmus or the lower part of the kidney. This is accomplished by nephropexy. After resection of the isthmus, the lower pole of the kidney is raised a little and moved laterally and then fastened by the capsule to the lumbar muscles with several sutures. A muscle graft can be used for the purpose. As a result the pelvis is moved medially and the ureter is positioned medially to the kidney, while the lower calyx is raised to the level of the prepelvic part of the ureter. Care should be taken, however, not to stretch the vessels in the kidney and not to cause circulatory disorders in it.

We did not think it always necessary to rotate the kidney to a position in which its pelvis faces medially, as is done by Foley (1940). We have sometimes encountered the vessels of the anomalous organ in an unusual position which approached its anterior and posterior surfaces. The kidney could not be rotated in such cases.

After plastic operations on the upper urinary tract we achieved drainage of the wound for seven to nine days and of the urinary tracts for two to three weeks by means of a perforated polyethylene drain.

Choice of operation. The diversity of the causes of the pathological conditions in fused kidneys and the frequently encountered poor results of surgical management have led to the use of fundamentally different operations for treating this anomaly and its diseases.

In the **treatment of disorders induced by a horseshoe kidney**

that is not complicated by disease, Martynov (1910) and other surgeons attempted to relieve the permanent pressure of the anomalous kidney on the large vessels and nerve plexuses by dividing or resecting the isthmus and, sometimes, performing nephropexy after that. We found descriptions of 47 such operations in the literature.

Fedorov (1923), on the contrary, believed that the results of isthmotomy were favourable because the kidney was fastened at the end of the operation and it was therefore quite sufficient to restrict the intervention to nephropexy. Twenty such operations are described in the literature available to us.

Attempts were also made to correct the disorders associated with fused kidneys by denervation (Hanley, 1942; Atanasiu et al., 1959).

Sokolov's operation (1930), comprising the resection of nervous anastomoses which intersect the isthmus of a horseshoe kidney, was not employed in the clinical practice because it was dangerous and not substantiated theoretically.

Proceeding from the fact that, besides pressure of the kidney on the large vessels and nerve plexuses, compression above the kidney leading to urinary stasis in the pelves is conducive to the development of disorders, we believe the first group of operations to be more expedient in these cases, because with division or excision of the isthmus better conditions are created for the drainage of urine from the pelves. In considering the indications in favour of this or that operation one should take into account the individual features of each case, i.e. the position of the pelvoureteral anastomosis, the localization of the accessory vessels, the thickness of the renal isthmus, and the causes of the disorder. It is usually possible to remove the causes of the disorders by resection of the isthmus of the horseshoe kidney and nephropexy or by resection of the isthmus with part of the lower pole of the kidney and nephropexy. The following case is given in illustration.

P., a 31-year-old woman, was admitted to the clinic on May 29, 1968. In the previous two years she had experienced frequent attacks of sharp pain in the right half of the abdomen, as a result of which she had become incapable of working. A painless, elastic, and poorly mobile tumour was palpated in the region of the umbilicus. Pasternatskii's and Gregoir's signs were positive. Blood and urine tests showed normal values. Indigo carmine was discharged from both ureteral orifices in the fourth minute. An image of a horseshoe kidney was found on a scout radiograph. The results of excretory urography confirmed the diagnosis of a horseshoe kidney and showed that it was not involved in any disease. An operation was carried out by the intraperitoneal approach. The isthmus of the horseshoe kidney, $1 \times 4 \times 2$ cm in size, was exposed and resected with part of the lower pole of the kidney to the level of the pelvis by means of the УКЛ-60 stitcher (Fig. 63). The lower pole of the kidney was then shifted a little laterally and fastened

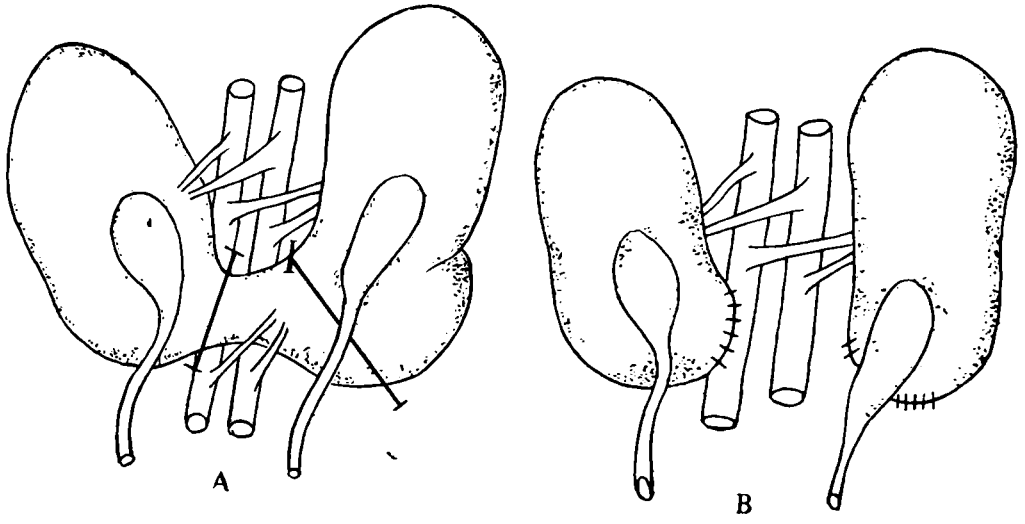


Fig. 63. Schematic representation of surgery on a horseshoe kidney with resection of the isthmus and part of the lower pole of the kidney and left nephropexy. *A*, before operation. *B*, after operation

by the capsule to the lumbar muscles with a catgut suture. As a result the prepelvic part of the ureter proved to be below the level of the lower pole. A drain was left in the wound. The postoperative period was uneventful.

After such an operation the prepelvic part of ureter is positioned at a level below that of the lower pole of the kidney and urine is adequately drained away from the pelvis. Some patients note pain on the other side due to mobility of the kidney. In such cases nephropexy is performed on this side four to six months after the first operation. To avoid the second operation, bilateral stage-by-stage nephropexy can be carried out during the first intervention.

Diseases of fused kidneys are most commonly managed by **heminephrectomy.** Eisendrath et al. (1925) found in the literature information on 71 such operations performed up to 1925. In 1955 Culp and Winterringer reported on 52 heminephrectomies carried out in one clinic. In the available literature we found reports on over 200 such operations performed to date. Heminephrectomy is usually undertaken in cases of large and multiple stones, stones in kidneys affected by tuberculosis, polycystic disease or tumour, in recurrent nephrolithiasis after pyelolithotomy and nephrolithotomy, in cases with hydronephrosis, pyelonephritis, trauma, tumour, tuberculosis or hydatid disease of the kidney, solitary cyst, profuse haematuria in polycystic fused kidney, etc.

The predisposition of fused kidneys to bilateral diseases, to disease in general, and to combinations of diseases calls for

a more conservative approach to the management of these conditions. That is why, as we find from our own experience, the indications for heminephrectomy should be limited. It can be recommended in cases of atrophy of the renal tissue and loss of function in one of the halves of the kidney. The following case serves as an illustration.

G., a 36-year-old man, was admitted to the clinic on November 17, 1966 complaining of permanent dull pain in the epigastrium that had troubled him for the last 12 years. On examination, an elastic, painless, and poorly mobile tumour was palpated to the left of the umbilicus and extended to the hypochondrium. Pasternatskii's sign was positive on the left. Blood tests showed normal values. Urinalysis: reaction alkaline, specific gravity 1018, protein 1.65 pro mille, leucocytes and erythrocytes cover the field of vision. Urine from the right kidney contains 2 to 4 leucocytes in field of vision, from the left kidney 85 to 90 leucocytes in field of vision. Urine culture produced growth of colibacillus. Indigo carmine was discharged from the right ureteral orifice in the fourth minute and had still not discharged at all from the left orifice after 15 minutes. A urogram showed the right anomalous pelvis on a level with the second lumbar vertebra; the left pelvis could not be seen. With retrograde pyelography the left pelvis was shown as the shadow of a large cavity situated on a level with the eleventh thoracic and third lumbar vertebrae. A diagnosis of left hydronephrosis and pyelonephritis of a horseshoe kidney was made. In an operation the left half of the horseshoe kidney was exposed together with a parenchymal isthmus that was $6 \times 3 \times 2$ cm in size. The isthmus was divided by means of the УКЛ-60 stitcher. Three vascular pedicles of the kidney and the ureter were ligated separately. The kidney was removed. It was found to be a multilayered fibrous sac with small areas of renal tissue at the isthmus.

Preservation of the hydronephrotic half of the horseshoe kidney was inexpedient in this case, since there was no functioning renal tissue. We undertook heminephrectomy for the same indications when the contralateral half of the kidney functioned well but was also affected with hydronephrosis. A few months after heminephrectomy these patients underwent plastic surgery on the remaining part of the kidney.

Heminephrectomy is indicated in hydronephrosis when localized purulent pyelonephritis or acute purulent infection develops. It is necessary in calculous pyonephrosis and calculous hydronephrosis with a non-functioning kidney. We preferred primary heminephrectomy in such cases to avoid the difficulties of reoperation on the fused organ, because the sclerotic process conceals the isthmus and the numerous accessory vessels even more and joins the horseshoe kidney to the aorta and vena cava.

The operation is conducted in cases of polycavitary tuberculosis of the fused kidney and pyonephrosis, when half of the kidney does not function. Preservation of the kidney in such cases is not called for.

Patients with tumour of half of the horseshoe kidney are also

subject to heminephrectomy. In localized tumours of the isthmus the latter is resected.

Reports are found in the literature of **operations, commonly performed for different diseases**, being carried out on fused kidneys. Pyelolithotomy is the preferred operation for nephrolithiasis. Partial nephrotomy is rarely conducted due to the danger of injuring the numerous blood vessels (Bogdansky, 1950). Some surgeons recommend repeating pyelolithotomy in recurrent nephrolithiasis (Gasparyan, 1962). The usual plastic operations are sometimes performed for hydronephrosis (Berman and Sidorenko, 1958), nephropexy for nephroptosis (Eliseev, 1968), resection of the cyst or kidney for a solitary cyst (Masyukova, 1949), extirpation of the cyst for hydatid disease (Viktorov et al., 1963).

It is our experience that the common organ-preserving operations which do not involve division of the kidney do not correct the unfavourable anatomical structure of the anomalous organ and therefore do not remove the causes of chronic urinary stasis in the renal pelves. They should be used only in the absence of marked dyskinetic disorders in the upper urinary tract and urinary stasis, but this is rarely encountered with this anomaly.

Reconstructive organ-preserving operations have not found wide use in the treatment of diseases of fused kidneys to date. Only a few cases of such operations are reported in the literature and operations of the same type are undertaken even with different causes of the disease.

In hydronephrosis some surgeons perform isthmotomy alone or in combination with nephropexy (Chikov and Rubtsov, 1964), or resect the isthmus of the horseshoe kidney and then carry out nephropexy (deBacker, 1961). Some surgeons perform plastic surgery on the upper urinary tract after such an operation (Shatin, 1963; Klyucharev and Petrova, 1966). In the available literature we found mention of 34 such operations. Besides, attempts are made to treat hydronephrosis with secondary calculosis by means of isthmotomy, pyelolithotomy, and plastic surgery of the pelvis and nephropexy (Gammelgaard, 1954; Culp and Winterringer, 1955). Gregoir (1963) believes that compression of the ureter on the isthmus and by accessory vessels can be relieved completely and the pelvoureteral anastomosis widened by simultaneous resection of the isthmus and lower pole of the horseshoe kidney and resection and plastics of the pelvis.

Our experience shows that in choosing the reconstructive operation the causes of the disorders should be considered individually in each case. Causes which may be linked with the structure of the renal anomaly should be taken into account and methods of their removal considered. With a routine approach to

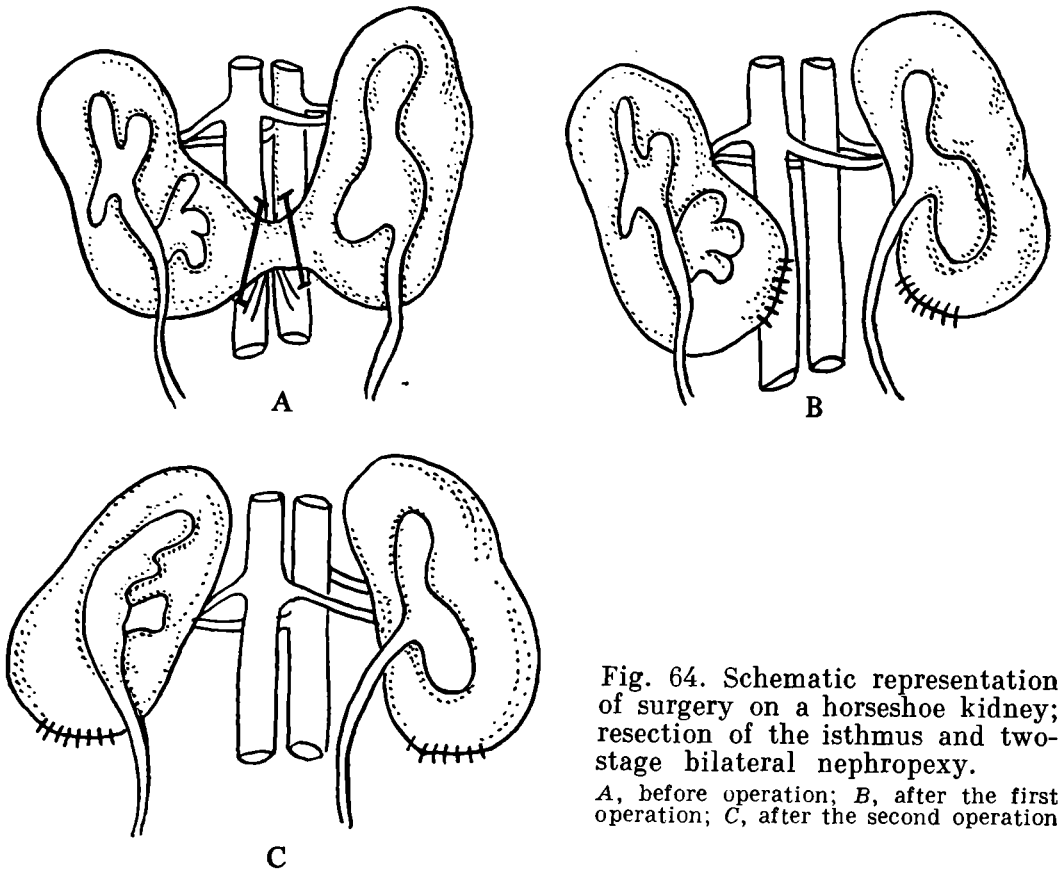


Fig. 64. Schematic representation of surgery on a horseshoe kidney; resection of the isthmus and two-stage bilateral nephropexy.
A, before operation; *B*, after the first operation; *C*, after the second operation

the choice of operation its outcome will be successful only by chance.

We performed several operations in our practice depending on the discovered cause of the disorder. In cases of a high origin of the ureter and its compression at the isthmus of the fused kidney we resected the isthmus and conducted nephropexy or removed the isthmus with part of the lower pole of the kidney (Fig. 64). As a result of the operation the prepelvic part of the ureter is now situated below the level of the pelvis, calyces and lower pole of the kidney. This creates good conditions for the drainage of urine.

In cases in which hydronephrosis developed due not only to the above-mentioned factors but also due to stricture of the ureter or its compression by the accessory vessels, a fixated ureteral kink or periureteritis, we performed, in addition to this operation, plastic procedures on the upper urinary tract. The combined operation allowed us to relieve compression of the ureter on the isthmus of the horseshoe kidney completely, widen the pelvoureteral anastomosis and free it from the pressure of scars and accessory vessels, shift the ureteropelvic segment to the

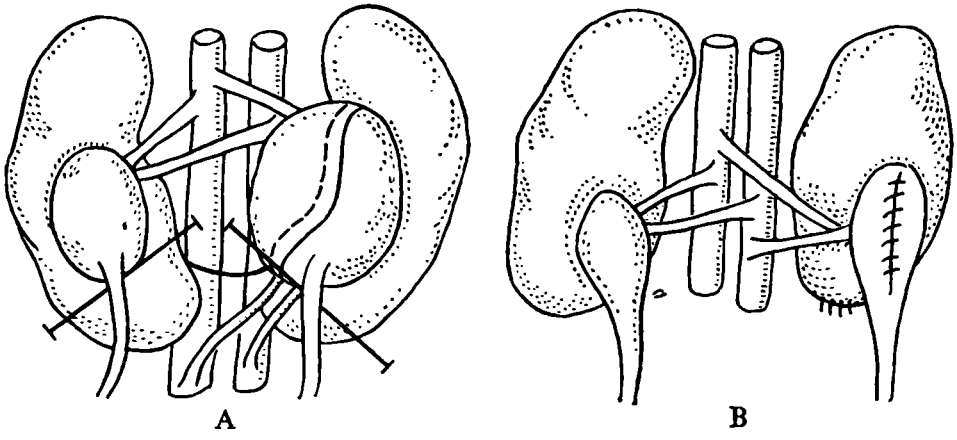


Fig. 65. Hydronephrosis of the left half of a horseshoe kidney. Diagram of operation for resection of the renal isthmus and pelvis and uretherolysis. *A*, before operation; *B*, after operation

lower part of the renal pelvis, lessen the size of the pelvis, and correct the vesicoureteral reflux. Depending on the circumstances, we used resection of the pelvis, ureterolysis, Lichtenberg's lateral pelvoureteral anastomosis, Foley's Y-plasty of the pelvis, ureteropyeloreimplantation and subtotal resection of the pelvis, excision of the main ureter of the duplicated half of a horseshoe kidney with the creation of anastomosis between the lower pelvis and the upper ureter, etc. The following case is given in illustration.

D., a 36-year-old man, was admitted to the clinic on March 27, 1963. He complained of attacks of sharp lumbar pain attended by elevation of temperature and a chill. He had been ill for the last 13 years. In 1960 a stone was removed from the right half of a horseshoe kidney. Examination one year after the operation revealed recurrent stones in the right half and hydronephrosis of the left half of the horseshoe kidney. In 1962 Fenger's operation was carried out on the left half of the kidney, after which the patient's condition became even worse. The attacks of sharp pain occurred more frequently and ceased only after catheterization of the left renal pelvis. On examination an elastic, painful, and poorly mobile tumour was palpated in the left hypochondrium. Pasternatskii's sign was positive on the left. After urological examination, the diagnosis of bilateral hydronephrosis and pyelonephritis of a horseshoe kidney and recurrent stones in the right half was made. During operation a parenchymal isthmus of the horseshoe kidney, $6 \times 4 \times 1$ cm in size, and the prepelvic part of the ureter were separated from gross scars. The ureteropelvic orifice was found at a high level. Resection of the isthmus and saddle resection of the pelvis were carried out (Fig. 65). After surgery the ureteropelvic segment was below the pole of the kidney. The postoperative course was uneventful. The patient recovered.

Resection of the pelvis in this case made it possible to transpose the ureteropelvic segment downward and remove part of the thick pelvic wall which could not contract spontaneously. Even

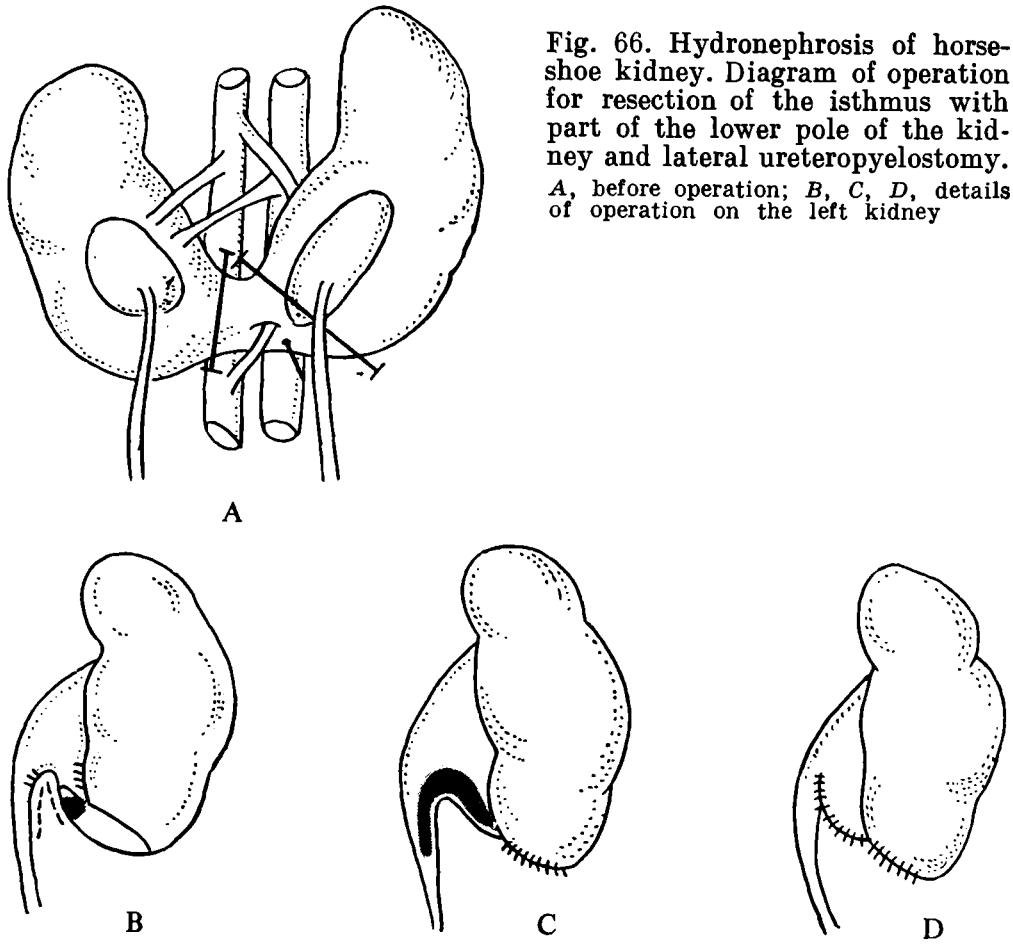


Fig. 66. Hydronephrosis of horseshoe kidney. Diagram of operation for resection of the isthmus with part of the lower pole of the kidney and lateral ureteropyelostomy. *A*, before operation; *B*, *C*, *D*, details of operation on the left kidney

this surgical intervention, however, could not create favourable conditions for drainage of the pelvis until the isthmus of the horseshoe kidney was resected. It should be noted that the plastic procedures earlier performed on this half of the anomalous organ did not hinder the operation.

With high origin of the ureter and its compression on the isthmus of the horseshoe kidney, good conditions for the flow of urine from the pelvis can be created by resection of the isthmus with part of the lower pole of the kidney, resection of the pelvis and nephropexy.

Cases of high origin of the ureter and compression of its prepelvic part by accessory vessels or a stricture of this part are successfully managed by isthmectomy or resection of the isthmus with the lower pole. In the first variant nephropexy is advisable.

Adequate conditions for the flow of urine from the pelvis are provided by resecting the isthmus with part of the lower pole

of the kidney and performing plastic procedures on the upper urinary tract. There will then be no need for nephropexy.

K., a 44-year-old woman, was admitted to the clinic on December 29, 1965. She complained of frequent attacks of sharp pain in the left lumbar region which were attended with vomiting, elevation of body temperature, chills, and frequent and painful micturition. For a period of four years she had felt dull pain in the small of the back between attacks. On the basis of urological examination the diagnosis of bilateral hydronephrosis and pyelonephritis of a horseshoe kidney was made. During operation it was established that the causes of hydronephrosis were a high origin of the ureter, stricture of its prepelvic segment and compression of this segment above the renal isthmus. The isthmus was resected together with part of the lower pole of the left half by means of the УКЛ-60 stitcher (Fig. 66). Part of the renal pelvis was also resected. The incision on the pelvis was continued on the ureter. Lichtenberg's ureteropelvic anastomosis was created. As a result of these manipulations the ureteropelvic segment was transposed to the lower corner of the pelvis. The postoperative period was uneventful. The patient recovered.

In the case described above the lower calyx involved in hydrocalycosis was resected together with the isthmus and lower pole of the kidney and the lower part of the renal pelvis was removed.

In some patients in this group we resected the isthmus with part of the lower pole of the horseshoe kidney and carried out Foley's Y-plasty with a triangular graft. When large pelves were involved in fibrosis we resected, in addition, part of the pelvis, which also promoted downward displacement of the ureteropelvic segment.

K., a 38-year-old man, was admitted to the clinic on April 6, 1964. He complained of dull pain in the epigastrium and both hypochondriac regions, frequent bilateral renal colic with vomiting, elevation of body temperature, a chill, and frequent and painful micturition. He had been ill for 14 years. An elastic, painless, and poorly mobile tumour was palpated in the region of the umbilicus. Pasternatskii's sign was positive on both sides. On the basis of urological examination the diagnosis of coral calculus in the right half and hydronephrosis of the left half of a horseshoe kidney and bilateral pyelonephritis was made. An operation was carried out on April 23, 1964 with a right lumbar approach. The isthmus of the horseshoe kidney, $4 \times 3 \times 1$ cm in size, was exposed and resected by means of the УКЛ-60 stitcher. The coral calculus was found in an extrarenal pelvis and extrarenal calyces. It was removed through an incision made in the pelvis and one of the calyces containing its most branching process. The incision in the pelvis and calyx was closed with interrupted catgut sutures. The postoperative period was uneventful. On June 11, 1964 the left kidney was exposed through a left lumbar incision. An enormous pelvis was found on the anterior surface of the kidney, the ureter originated at a high level and its prepelvic part was compressed by scars above the lower pole of the kidney. The lower pole was resected by means of the УКЛ-60 stitcher (Fig. 67). Foley's Y-plasty of the pelvis with an inferior graft was then performed. As a result of this operation the prepelvic segment of the ureter took a position below the pole of the kidney. The postoperative period was uneventful. The patient recovered.

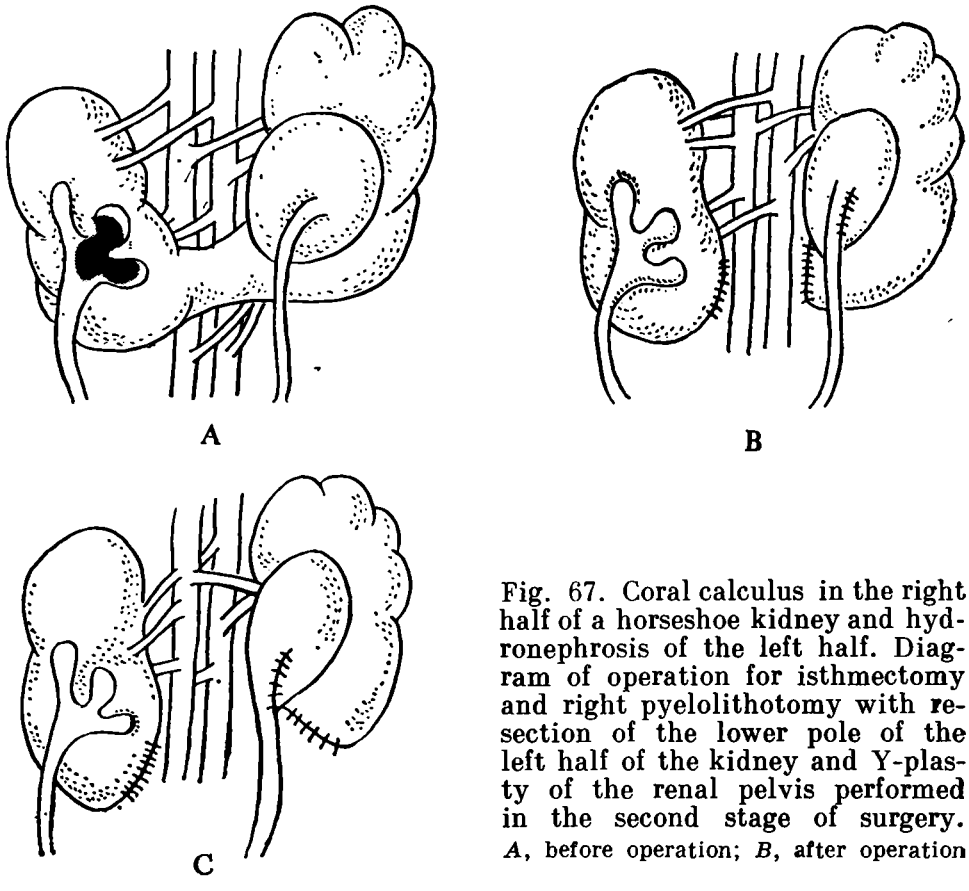


Fig. 67. Coral calculus in the right half of a horseshoe kidney and hydronephrosis of the left half. Diagram of operation for isthmectomy and right pyelolithotomy with resection of the lower pole of the left half of the kidney and Y-plasty of the renal pelvis performed in the second stage of surgery. *A*, before operation; *B*, after operation

The purpose of surgery in this case was to remove the cause of the developing renal insufficiency, namely the coral calculus on the right side and hydronephrosis of the left half of the horseshoe kidney.

In cases of high origin of the ureter and fixed kinking we used subtotal resection of the pelvis and reimplantation of the ureter and pelvis, as in the following case. It should be stressed that the patient had been subjected earlier to heminephrectomy and the subsequent intervention was carried out on the remaining half of the horseshoe kidney.

Kh., a 39-year-old woman, was admitted to the clinic on February 4, 1964. She complained of dull pain in the right hypochondrium and right lumbar region which often became sharp and was then attended with elevation of body temperature, a chill, a frequent and painful micturition. She had had the dull pain since childhood. She underwent appendectomy which brought no relief and was admitted many times to a surgical clinic because ileus was suspected. In 1962 she underwent heminephrectomy of the left half of a horseshoe kidney for calculous hydronephrosis. It was during this operation that the horseshoe kidney was discovered. After surgery Kh. was still troubled by pain in the right half of the abdomen and right lumbar region, and now and then experienced a rise in temperature and a chill.

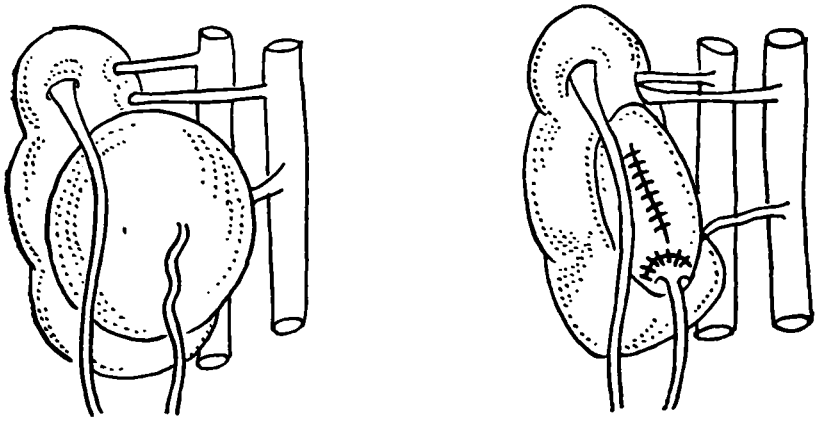


Fig. 68. Hydronephrosis of the lower part of a duplicated solitary right half of a horseshoe kidney remaining after left heminephrectomy. Diagram of operation for subtotal resection of the renal pelvis and reimplantation of the ureter into the pelvis.

A, before operation; *B*, after operation

It was recommended that Kh. should take up residence in some other climate. On admission to the clinic arterial pressure was 140/80 mm Hg. An elastic, painless, and poorly mobile tumour was palpated in the umbilical region. Pasternatskii's sign was positive on the right. Blood test: Hb 78 per cent; leucocytes 8700: eosinophils 3, rod neutrophils 5, filamented neutrophils 70, lymphocytes 16, monocytes 6; ESR 18 mm/hour. Urinalysis: reaction acid, specific gravity 1020, protein 0.66 pro mille, leucocytes and erythrocytes covering the entire field of vision. Urine culture produced growth of staphylococcus. Indigo carmine was discharged from the right ureteral orifice in the third minute. From the findings of urological X-ray examination the following diagnosis was made: hydronephrosis and pyelonephritis of the lower half of the right duplicate kidney remaining after resection of the left half of a horseshoe kidney. It was found during surgery that the lower two thirds of the right half of the horseshoe kidney were a large hydronephrosis with an enormous pelvis, the prepelvic segment of the ureter was curved and fastened to the pelvis with scars (Fig. 68).

The upper part of this kidney had a small pelvis whose ureter curved over the front surface of the hydronephrosis. The prepelvic ureteral segment was freed from the scars and cut off, together with a pelvic rosette. Subtotal resection of the hydronephrotic pelvis was then performed. Its edges were sutured and the pelvic rosette was stitched into the lower corner. The postoperative period was uneventful. The patient recovered.

With compression of the prepelvic part of the ureter on the isthmus of the horseshoe kidney and reflux into the main ureter of its duplicate half, we removed the causes of the disease by resecting the isthmus with part of the lower pole of the horseshoe kidney, excising the main ureter, creating anastomosis between the lower pelvis and the upper ureter, and by performing nephropexy.

V., a 20-year-old woman, was admitted to the clinic on January 4, 1968 complaining of frequent attacks of sharp pain in the left lumbar region attended with a rise in body temperature, a chill, and frequent and painful micturition. In the periods clear of attacks she felt dull pain in the small of the back, at times pulsation in the abdomen and paraesthesia of the lower limbs. She had been ill for three years. Non-operative treatment was ineffective. Because of all these disorders V. had had to give up work. Examination revealed an elastic, painless, and poorly mobile tumour in the umbilical region. Pasternatskii's and Gregoir's signs were positive. A blood test showed normal values. Urinalysis: reaction acid, specific gravity 1019, protein 0.99 pro mille, leucocytes covering the entire field of vision, erythrocytes 25 to 30 in field of vision. Urine culture produced growth of colibacillus. In the urinary bladder there were one ureteral orifice on the right and two orifices on the left. Indigo carmine was discharged from the right and lower left orifices in the fourth minute and from the upper left orifice in the seventh minute. Urine from this pelvis contained 25 to 30 leucocytes in field of vision. On the basis of urological X-ray examination the following diagnosis was made: hydronephrosis and pyelonephritis of the left lower double half of a horseshoe kidney, reflux into the main ureter on the left. During operation a parenchymal isthmus of the horseshoe kidney $2 \times 3 \times 1$ cm in size was exposed and resected with part of the lower pole on the left by means of the УКЛ-60 stitcher. The diagram of the operation is shown in Fig. 69.

The ureter of the lower pelvis was excised. A lateral anastomosis 2.5 cm long was formed between the lower pelvis and the upper ureter by means of a

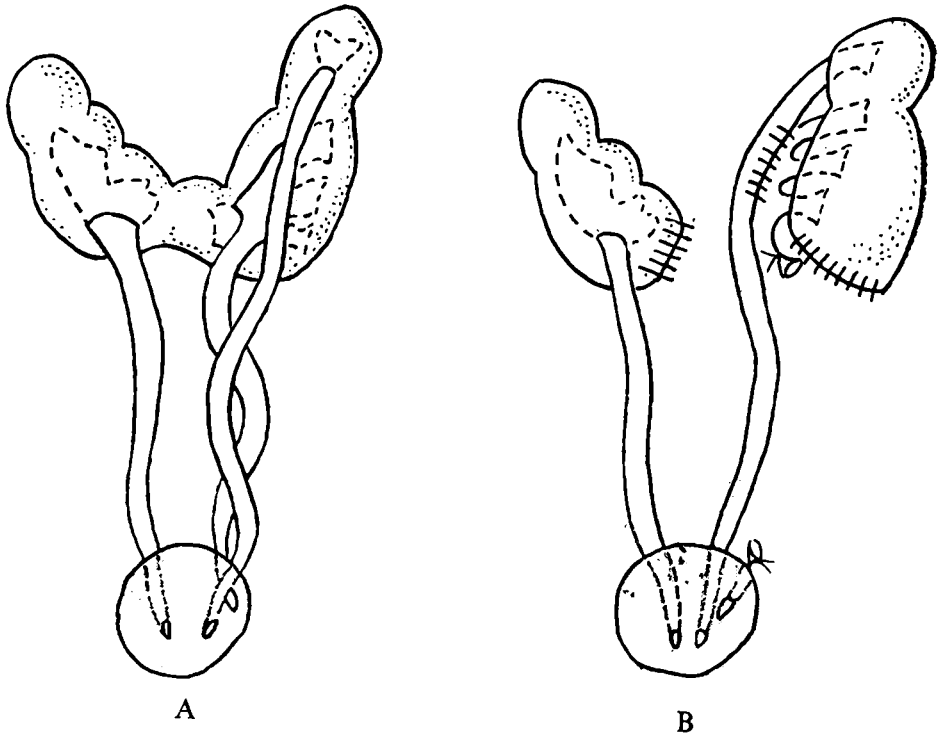


Fig. 69. Hydronephrosis and pyelonephritis of the left duplicated half of a horseshoe kidney. Diagram of operation for resection of the isthmus and part of the lower pole of the left half of the horseshoe kidney, resection of the main ureter and ureteroneopyelostomy.

A, before operation; *B*, after operation

single row of interrupted catgut sutures. The calyces of the lower pelvis were extrarenal. The lower pole of the kidney was shifted a little laterally and fastened by the capsule to the lumbar muscles with a catgut suture. The postoperative period was uneventful. The patient recovered.

Thus, various reconstructive operations are used in hydronephrosis of horseshoe kidneys depending on the different anatomical conditions and causes of the disease. In cases when the disease has not advanced far it can be expected that the operations will restore the function of the kidney and bring about favourable morphological changes in it. In other cases they will bring relief to the patient and prevent further destruction of the kidney and disturbance of its function, i.e. the operations are among the measures of controlling the further development of renal insufficiency.

In **nephrolithiasis** with no marked hydronephrosis of the horseshoe kidney it is necessary to remove the obstacle to the flow of urine in the prepelvic segment of the ureter (high origin of the ureter, compression of the ureter above the renal isthmus, etc.). For this purpose the isthmus of the anomalous organ is resected (if possible together with part of the lower pole of the kidney), pyelolithotomy is carried out, and whenever necessary nephropexy is also performed. Nephropexy in this case also involves shifting the lower pole of the kidney laterally and fixing it in this position.

In cases of marked hydronephrosis with secondary calculosis, the removal of the other causes of hydronephrosis by means of a plastic operation is expedient.

In **pyelonephritis** of a horseshoe kidney with severe urinary stasis in the pelvis a reconstructive operation may also be of value. It is usually performed for removing the obstacle to the flow of urine in the prepelvic segment of the ureter, that is, when it is compressed above the renal isthmus or the ureter has a high origin. In such cases we resected the isthmus of the horseshoe kidney with part of its lower pole and carried out nephropexy. Markin (1960) and Bazhenov (1961) pointed to the efficacy of isthmotomy with nephropexy or isthmectomy in pyelonephritis of a horseshoe kidney.

Results of surgical treatment. Judging from the data in the literature, heminephrectomy and simple organ-preserving operations on fused kidneys produce favourable immediate and early results. The late results of such operations, however, are very disappointing. There are descriptions in the literature of affections of the remaining part of the kidney after heminephrectomy, and of a recurrence of the disease after simple organ-preserving operations because the conditions for the development of the disease remain. Culp and Winterringer (1955), for instance, not-

ed recurrences in 50 per cent of patients who were operated on for nephrolithiasis and in two of eight patients with hydronephrosis. According to these surgeons, the average survival time of such patients after surgery is 11.5 years. Many patients die from renal insufficiency.

Most reconstructive organ-preserving operations also produce favourable immediate and early results. But the late results of such operations are described in only a few works and only a small number of cases are discussed. Moreover, the information in the literature on this problem is controversial. With disorders and the absence of diseases Foley (1940) gained good results with reconstructive operations. Culp and Winterringer (1955) note unfavourable outcomes in two of their cases. Some authors report that a urinary fistula which developed after reconstructive surgery called for another heminephrectomy. Shatin (1963) and Gregoir (1963) described good results of reconstructive operations in cases of hydronephrosis, but in cases reported by Nation (1945) and Deuticke(1962) further development of the disease required heminephrectomy to be repeated. Chikov and Rubtsov (1964) failed to achieve good results in three such operations. With nephrolithiasis Culp and Winterringer (1955) had good results but Foley (1940) reported a recurrence of the disease.

Analysis of the outcomes of the surgical management of the renal anomaly and its diseases shows the expediency to distinguish three groups: (1) the results of heminephrectomy, (2) of routine operations in which only the cause of the disease or its complication is removed, and (3) of reconstructive operations in which the consequences of the disease and renal anomaly are removed.

We assessed the postoperative results in our cases as follows: excellent (the patient has no complaints, his working capacity has been restored, the urine is normal, renal function is completely restored, normal size of the pelvis and calyces is restored, and there are no calculi); good (the patient has no complaints, his working capacity has been restored, there is mild pyuria, improved renal function, diminished size of pelvis and calyces); fair (subjective improvement, pyuria, no changes in renal function, arrest of kidney destruction); poor (complaints and pyuria remain, renal function deteriorates progressively, the pelvis and calyces increase in size, attacks of pyelonephritis occur, and there is recurrence of urolithiasis).

It is our experience that the final results of operations on fused kidneys are usually evident within the first two years after the intervention. This mainly applies to the management of retention diseases, i.e. hydronephrosis, nephrolithiasis and pyelone-

phritis. The late results should therefore be judged no earlier than this period.

We studied the results of 92 operations on fused kidneys of 68 patients. They are shown in Table 4.

Table 4. Results of Operations on Horseshoe Kidneys

Operation	Number of cases	Results			
		Excellent	Good	Fair	Poor
Heminephrectomy	19	4	8	4	3
Simple organ-preserving operations	33	5	8	3	17
Reconstructive organ-preserving operations with resection of the isthmus and pole of the horseshoe kidney	40	12	17	7	4
Total	92	21	33	14	24

The results of heminephrectomy for hydronephrosis, nephrolithiasis, tuberculosis and tumours of horseshoe kidneys were studied in 19 of our patients in follow-up periods of two to eleven years. They were good in twelve patients, fair in four and poor in three. The poor outcomes were linked with the development of hydronephrosis or calculus remaining in the kidney after heminephrectomy or with metastasis of hypernephroma. The fair results were due to the development of the initial signs of diseases of the solitary kidney. These are also potentially poor results.

The outcomes of simple organ-preserving operations without isthmotomy were studied in follow-up periods of 2 to 16 years. They were poor in most cases (in 17 of 33).

We encountered poor results for ureterolysis and when ureterolysis was combined with Fenger's operation. In one of these cases heminephrectomy had to be undertaken because of localized purulent pyelonephritis, while in another case a reconstructive operation was repeated on this kidney.

Poor results are noted particularly often with pyelolithotomy, nephrolithotomy and ureterolithotomy. Recurrences of nephrolithiasis were revealed in 14 cases. One patient died from progressive renal insufficiency associated with developing hydronephrosis and pyelonephritis. The poor outcomes of such operations are evidently caused by the persisting stasis of urine in the pel-

ves (due to the developmental anomaly of the organ) which had not been corrected during the operation.

In the other cases in which there was no urinary stasis in the pelves the results were excellent, good or fair. Under such conditions evolution of the process is particularly conspicuous in children.

Only fair results were achieved by nephropexy of nephroptosis in cake and unilateral fused kidneys. They brought temporary relief from pain for one or two years, after which the pain disorders recurred.

The results of reconstructive operations with resection of the isthmus of the horseshoe kidney were studied in follow-up periods of three to eleven years. They were poor in four of forty cases.

Resection of the isthmus with part of the pole of the kidney and nephropexy were performed in all cases, plastic procedures on the upper urinary tract were carried out in 21 cases, and pyelolithotomy in 15. Nephropexy was not an independent operation in these cases, but only a component of the reconstructive intervention.

This operation proved effective in dyskinesia of the upper urinary tract, hydronephrosis, nephrolithiasis, pyelonephritis, nephroptosis and tumour of the isthmus of the horseshoe kidney, when it was necessary to remove different causes of urinary stasis in the upper urinary tract. Excellent and good results were noted in 29 cases.

In seven patients the results were fair. These were cases with stage III hydronephrosis with markedly diminished renal function, when heminephrectomy could not be performed in a bilateral disease and renal insufficiency and the aim of the surgery was to improve the flow of urine from the pelvis and prolong the patient's life.

Poor results of such operations were noted in stage II and III hydronephrosis. The poor outcomes were due to progression of pyelonephritis, recurrences of nephrolithiasis, inadequate choice of operation, and errors in plastic procedures.

All patients who underwent reconstructive operations are kept under out-patient medical surveillance. Only one female patient died eight years after resection of the isthmus of a horseshoe kidney for hypernephroma. Death occurred at an old age after pneumonia. Thus, the results of reconstructive organ-preserving operations on fused kidneys are better than those of heminephrectomy and simple organ-preserving operations. With a proper choice of the reconstructive procedures and early performance of the operation before considerable morphological and functional disorders develop in the anomalous kidney, these operations merit application in clinical practice.

CHAPTER IV

Kidney Dystopia

AETIOLOGY AND PATHOGENESIS

Kidney dystopia is due to the arrest of the development of the kidney bud during its migration from the pelvis to the lumbar region. In addition to the general causes discussed in Chapter I which are conducive to the development of an anomalous kidney, the persisting embryonal vessels preventing upward migration of the kidney also play a role in the development of various types of kidney dystopia (Alekseev, 1926). This, however, may be of secondary character.

Some authors have considered a low anomalous position of the kidney to be an atavistic sign. Alekseev (1935) contended that upward migration of the kidneys is prevented by the excessive pressure of the coil of the growing intestinal loops.

The size of the organ is probably of some importance in the development of its dystopia. A large kidney may meet with a greater number of obstacles in its upward migration. According to Bremer (1957), during upward migration the kidney abuts against the suprarenal gland in the eighth week of embryonal life; in a human embryo this gland is very large. The diaphragm is also an obstacle to migration. In agenesia of the suprarenals the kidney may move higher and even enter the thoracic cavity through the posterior opening in the diaphragm.

The arrest of the upward migration of the kidney usually leads to the retardation of its rotation. That is why the pelvis of a dystopic kidney often faces the front.

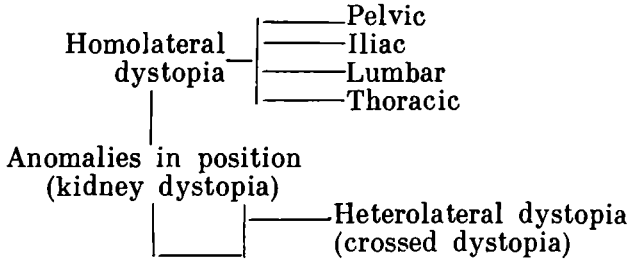
CLASSIFICATION

Dystopia, or malposition of the kidney, occurs when the kidney fails to migrate to the lumbar region during embryonal development and does not rotate about its axis. Such kidneys cease developing and, as distinct from nephroptosis, they never take up a position in the lumbar region at the normal level.

Depending on their position dystopic kidneys may be pelvic, iliac, lumbar or thoracic. Homolateral and heterolateral kidney dystopias are distinguished. In homolateral dystopia the kidney is found on the normal side, whereas in heterolateral dystopia it is displaced to the contralateral side of the body.

This anomaly may be unilateral or bilateral. In the bilateral anomaly the kidneys may be situated symmetrically or asymmetrically. Pelvic dystopia of both kidneys is an example of the symmetrical anomaly, while lumbar dystopia of the kidney on one side and iliac dystopia on the other are an example of asymmetrical malposition.

We believe that from the standpoint of the genesis of renal anomalies all positional anomalies related to fusion of kidneys should be included in the group of interrelationship anomalies, i.e. fused kidneys. In the group of positional anomalies we consider only dystopic kidneys which are not fused to each other. Thus, a diagram of this group of anomalies can be presented as follows



SURGICAL ANATOMY

The flat shape of a dystopic kidney, noted in most cases, is an embryonal feature. It is also often lobate. Its surface is irregular, often tuberous. There are depressions on its posterior surface corresponding to the vertebrae, pelvic bones and large vessels.

When in a high lumbar position the kidney sometimes has a normal bean-like shape. The lower its position, the greater the deviation from a normal shape. When in the small pelvis the kidney is often just a shapeless mass of renal tissue. The mechanical pressure exerted by the adjacent organs during development is evidently an important factor in the unusual shape of the dystopic kidney.

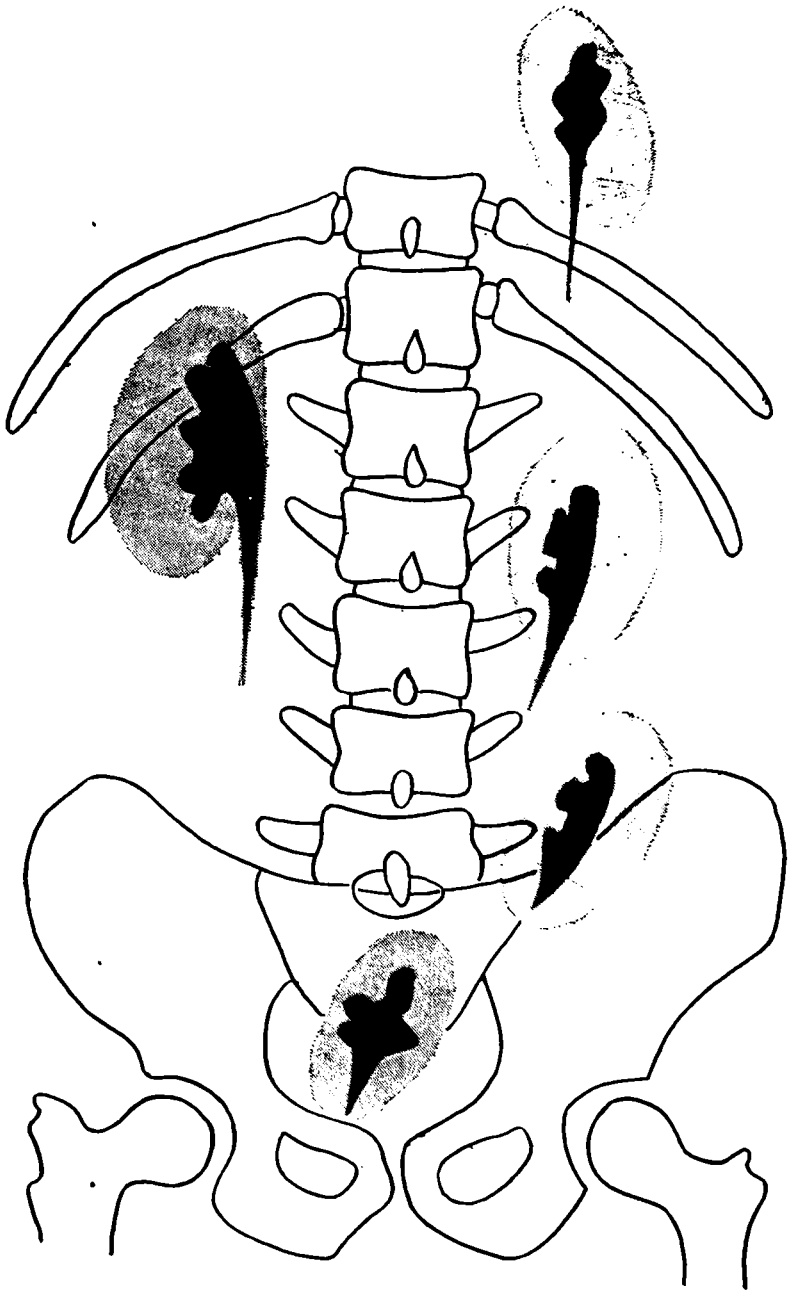


Fig. 70. Schematic representation of the main types of dystopic kidneys

According to Dretler et al. (1971), a dystopic kidney is always smaller than a normal one. Its average weight is 63 g. With lumbar dystopia, however, a kidney of normal size may sometimes be encountered.

If a dystopic kidney is not involved in a pathological process the structure of its parenchyma is normal. In terms of function it is also no different from normally developed kidneys.

The position of the kidney is one of the principal signs characterizing this renal anomaly. A dystopic kidney can be found in the pelvis (pelvic renal dystopia), in the iliac region (iliac dystopia), in the lumbar region below the normal level (lumbar dystopia), and above the diaphragm in the thoracic cavity (thoracic dystopia) (Fig. 70).

In crossed dystopia both kidneys are found on the same side of the midline of the body. The dystopic kidney in this case is usually below the contralateral kidney. This has been noted in the majority of cases by various authors (Pathak and Renjhen, 1972).

A dystopic kidney lies closer to the midline and. what is particularly important, usually lies closer to the anterior abdominal wall and is not protected by the ribs. This is why such a kidney is more vulnerable to injury.

The urinary tracts of dystopic kidneys are unusual. The hili are not always clearly defined. They may appear as a small depression or narrow fissure but sometimes they are quite normal in shape. In most cases they are found on the anterior surface of the organ, less frequently on the medial or lateral border, and very rarely on the posterior surface, as noted by Sigal (1934). The pelves are usually extrarenal but in rare cases they may be intrarenal. In four of our cases we found two pelves in the dystopic kidney, while the contralateral kidney was normal. Some authors distinguish four types of position of the renal pelvis in relation to the parenchyma and the midline of the body (Fig. 71).

In most cases the calyces of a dystopic kidney lie medial to the pelvis because the rotation of the anomalous kidney was not completed during its upward migration. In two of our cases of lumbar dystopia of the kidneys, however, they were lateral to the pelvis. The calyces are usually more numerous and branching. Extrarenal calyces are quite a frequent occurrence in this anomaly. We encountered them in nine operations. Komarovich (1957) and Fityo (1960) describe cases of extrarenal pelves with five extrarenal calyces.

In thoracic dystopia of the kidney the ureter is much longer than usual. In other cases it is shorter. The ureter originates from the lateral or middle part of the renal pelvis and usually lies above the pole of the kidney. The high and sharp-angled origin of the ureter and its position above the kidney are not conducive to good drainage of the pelvis; therefore, dystopic kidneys are predisposed to diseases in which urinary stasis plays a definite role.

The ureters terminate at the usual sites. In two cases in mutually crossed dystopia of the kidneys (Bugbee and Losee, 1919) each kidney was on the side contralateral to the orifice of its ureter in the bladder and the ureters consequently intersected one an-

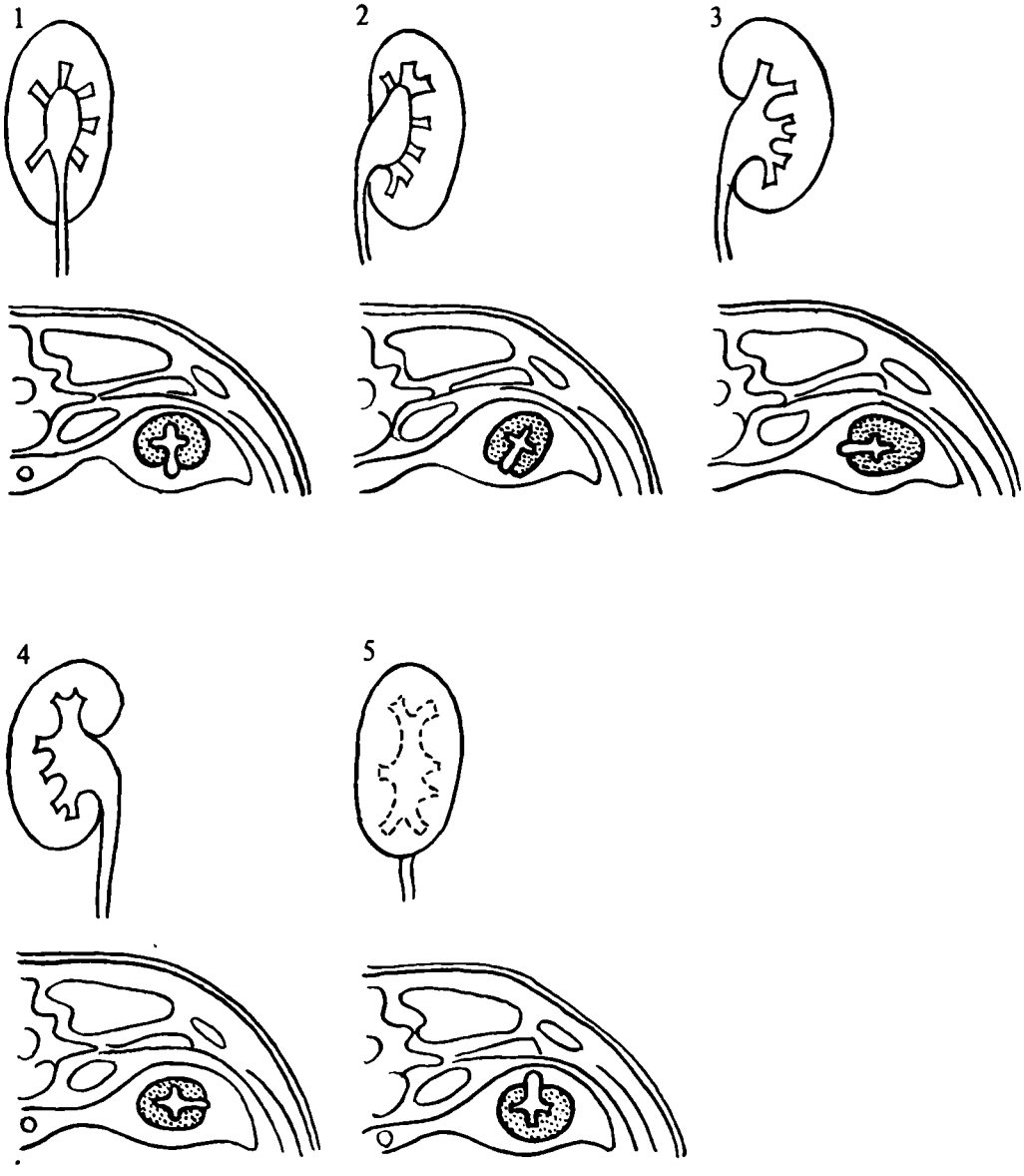


Fig. 71. Types of position of the pelvis of a dystopic kidney (after H. M. Wayrauch, 1939)

1, non-rotated kidney; *2*, incomplete rotation of the kidney; *3*, normal rotation of the kidney; *4*, reserve rotation of the kidney; *5*, reverse rotation of the kidney

other. But in other instances no peculiarities were noted in the position of the ureters.

Vascularization of the dystopic kidneys is distinguished by the variety in the course and number of vessels.

In most cases the number of vessels is considerably greater, and the lower the position of the anomalous kidney the more vessels it contains. Dretler et al. (1971) reported that in 17 of

33 kidneys there was only one artery, in 12 there were two arteries, three kidneys had three arteries and one kidney had four arteries. Shakhbazyan (1931) found six arteries in one dystopic kidney. In most of our surgical cases we found two or three arteries in one such kidney. This rich vascularization is explained by incomplete reduction of the numerous vessels of the mesonephros due to its incomplete embryonal development.

The arteries of a dystopic kidney branch from the aorta and the iliac and median sacral arteries at different levels. In addition, the subdiaphragmatic or mesenteric artery contributes to the blood supply of the kidney. Anichkov (1912) distinguished, on the basis of autopsies, the following four types of vascularization of dystopic kidneys: one artery originating from the bifurcation of the aorta, which is rarely encountered; numerous vessels arising from the aorta; numerous vessels arising from the aorta and large vessels of the pelvis; numerous vessels arising only from the large vessels of the pelvis, which is a rare occurrence. The main vessels arising from the aorta or iliac vessels enter the renal hilus. All the other vessels enter different parts of the kidney on the posterior or anterior surface, both close to the hilus and at a distance from it, at the poles. In some cases it is seen that the vessels directly penetrate the renal parenchyma.

The veins usually correspond to the number of arteries and are attendant to them. They drain into the vena cava inferior or its large branches.

These anomalies of the renal vessels can often be the cause of urinary stasis and diseases. They may create considerable technical difficulties during surgical intervention.

No special study has been made of innervation of dystopic kidneys. It is believed that this is accomplished by nerves branching from the same plexuses as in normally developed kidneys. This also applies to the lymphatic system.

A dystopic kidney is surrounded by a deficiently developed fat capsule owing to which it is poorly mobile. The numerous vessels also contribute to the lack of mobility. Due to its malposition such a kidney causes displacement of adjacent organs. Gruber (1866) described a case of abnormal position of the sigmoid colon. It was located transversely, reaching the right iliac region and passing from there into the minor pelvis. In pelvic dystopia of the kidney the urinary bladder may be displaced laterally. The iliac vessels are also shifted laterally. In some cases the suprarenal gland is either located at a low level next to the kidney or does not exist at all. In a case reported by Kokh and Kupershlyak (1932) the caecum was under the liver.

Most intrathoracic kidney dystopias are associated with anomalies of the diaphragm, congenital or acquired hernias. In some

cases a normally developed diaphragm has a small opening for the renal hilus (Zhuravlev and Shpanko, 1968).

There are several reports of occasional cases in which a dystopic kidney is combined with developmental anomalies of the kidney or other organs.

The urinary and genital systems develop from a common bud. That is why anomalies of the kidneys and genital organs are a relatively frequent occurrence. Katsyf and Khotman (1966) described hypoplasia of a pelvic dystopic kidney. Topchibashev (1957) had a case of a hypoplastic and duplicate dystopic kidney. Sucin and Caluser (1966) encountered a congenital solitary hypoplastic and dystopic kidney. Mentus (1968) had a patient with pelvic dystopia of a solitary kidney. Kawamura and Takahashi (1972) described a case of crossed dystopia of a solitary kidney. Among 42 autopsies Sokolov (1928) found two cases of a solitary dystopic kidney.

Occasional cases of anomalies of the genital organs and dystopic kidneys can be found in the literature in many observations. Dystopic kidneys and infantile genitals, hypo- and epispadias, cryptorchidism, high position of the ovary, agenesis of the deferent duct (Lurie et al., 1971), imperforate hymen, agenesis of the vagina, underdevelopment or agenesis of the uterus and its appendages, uterus bicornus (Petkevich, 1914; Donskoi, 1963), and duplication of the uterus and its appendages (Vainshtein, 1936) have been described. In our experience there were five cases of agenesis of the vagina, underdevelopment of the uterus and its appendages, agenesis of the uterine appendages, and cryptorchidism. In 13 of 97 cases in the practice of Thompson and Pace (1937) genital anomalies were combined with a dystopic kidney.

The literature has also recorded cases of dystopic kidney in conjunction with anomalies of other organs such as cleft palate, cleft lip, congenital underdevelopment of the suprarenal gland, fused suprarenals, incomplete development or agenesis of the anus, infantile or flat pelvis, spina bifida (Shakhbazyan, 1931; Malek et al., 1972). The combination of these anomalies with a dystopic kidney may be explained by the high incidence of developmental anomalies in general. It should be noted that such combinations are discovered at autopsy more often than they are encountered in clinical practice. This suggests that the total number of such combinations is actually greater than the number that has been described.

Statistics. Judging from the data in the literature and our experience dystopic kidneys are encountered quite often. Gerasimov and Vzorov (1931) collected information on 167 dystopic kidneys in the literature published up to 1930. In the literature

published in the Soviet Union and abroad to date we have found reports on about 2000 cases.

Most reports in the literature deal with individual cases, but with years some clinics have accumulated more statistical data. Thompson and Pace (1937) provide information on 97 cases, Akhundov (1968) on 98 cases, Eliseev (1968) on 83, Rabinovich (1968) on 82, Dretler et al. (1971) on 86 cases. Ostropolskaya and Bogdanova (1967) report on 38 dystopic kidneys in children. Over a period of 19 years (1956-1974) we had 162 patients with dystopic kidneys.

The incidence of dystopic kidneys found at autopsy, according to various researchers studying this problem on the basis of large number of cases, is approximately the same. Sokolov (1928) encountered dystopic kidneys in 0.08 per cent of autopsies. The same figure is cited by Thompson and Pace (1937) and Dretler et al. (1971). Anomalous position of the kidneys is encountered more frequently in clinical practice.

Among all the patients of their urological clinic Smith and Orkin (1945) found that 0.1 per cent had dystopic kidneys. Among patients with diseases of the kidneys and upper urinary tract Pekarskaya (1955) found this anomaly in 1.7 per cent, Tarasov (1963) in 0.9 per cent and Eliseev (1968) in 0.5 per cent. Goloshchapov (1969) noted dystopic kidneys in 1.9 per cent of children with diseases of the kidneys and upper urinary tract. Among our urological patients 0.11 per cent had dystopic kidneys, while among those with diseases of the kidneys and upper urinary tract 1.0 per cent. During surgery we discovered dystopic kidneys in 1.8 per cent of patients. According to Deess (1941) dystopic kidneys were found in 0.2 per cent of patients subjected to X-ray examination, our data, 0.4 per cent, are the same as those reported by Culp and Hiebert (1944) and Vourinen (1962).

This discrepancy in autopsy findings and clinical data points to the fact that many dystopic kidneys have no clinical manifestations and remain undiscovered.

There is a difference of opinion with regard to the predisposition of males or females to this anomaly. According to Khrushchov (1962), it is prevalent among males, but most other researchers contend that dystopic kidneys are discovered more frequently in females (Gimpelson, 1949; Raevsky, 1957; Derevyanko, 1968). Females were also in a majority among our cases (93 females, 69 males). The higher incidence among women is evidently explained by the fact that the anomaly is discovered more frequently in diseases of the female genitals and during pregnancy. Significance in this regard should be attached to the total number of males and females treated at the clinic. According to Sokolov

(1928), there are more female patients at these clinics. We also find this to be true.

Dystopic kidneys are found predominantly in patients aged between 20 and 50 years (Gimpelson, 1949; Raevsky, 1957; Derevyanko, 1968). Most of our patients with dystopic kidneys were also of this age group. In children this anomaly is found most frequently at the age of 7 to 15 years (Goloshchapov, 1969). However, the anomaly may be encountered at any age from early childhood till old age. The large number of cases revealed among the middle-aged is evidently associated with the fact that at this time of life the organism is exposed to maximum effort and the various anomalies become apparent. In children this anomaly is often revealed in the clinic in association with concomitant developmental anomalies of different organs and their complications.

In our experience, dystopic kidneys rate third in frequency among the various renal anomalies. Dystopic kidneys account for 11.5 per cent (162 cases) of 1405 anomalies. In the experience of Plotnikov (1966) and Antonov (1968) they accounted for 8.5 to 16.2 per cent of cases; Gimpelson (1949) and Akhundov (1968) found them in 20.4 to 27.8 per cent, and Raevsky (1957) in 40.4 per cent of cases.

In clinical practice lumbar dystopia of kidneys is encountered more often, iliac dystopia is less frequent, while pelvic or thoracic dystopia is even rarer. Crossed dystopic kidneys and bilateral kidney dystopia are also rare. Among our patients 76 had lumbar dystopia, 43 had iliac dystopia, 14 had pelvic dystopia, 7 had crossed dystopia and 12 had bilateral dystopia. According to Gimpelson (1949), lumbar dystopia was found in 28 patients, iliac in 6 and pelvic in 7. Ostropolskaya and Bogdanova (1967) found lumbar dystopia in 24 patients, iliac in 10, pelvic in 1 and thoracic dystopia in 3 patients.

Toole et al. (1961) point out that no more than 50 cases of intrathoracic dystopia of the kidneys are reported in the literature. In later years only occasional cases are reported.

Berg and Kearns (1949) cite information found in the literature on 45 pelvic solitary kidneys. In the Soviet literature Vainshtein (1936) and Kantariya (1938) each reported one case of solitary pelvic kidneys.

Lee (1949) found reports in the literature describing 29 cases of crossed renal dystopia. Dworin (1969) encountered one very rare case of crossed dystopia of a solitary kidney.

Coppridge (1934) provides summarized statistics on 22 cases of bilateral pelvic kidney dystopia.

Dystopic kidneys are most frequently found on the right side. Among our 133 patients with homolateral dystopia 81 had the

anomaly on the right side and 52 on the left. Among 12 patients with bilateral dystopia six had bilateral lumbar dystopia, one had lumbar dystopia on the right and iliac dystopia on the left, and five patients had a dystopic lumbar kidney on the left and an iliac dystopia on the right. Thompson and Pace (1937) and Gimpelson (1949) also pointed to the high frequency of this renal anomaly on the right side. Dretler et al. (1971) contend that this anomaly occurs on both sides with equal frequency. According to the autopsy findings made by Sokolov (1928), 0.042 per cent of cases were left dystopia, 0.038 per cent right dystopia and 0.004 per cent bilateral dystopia.

The diseases of dystopic kidneys have not been sufficiently studied. Akhundov (1968) reported that nephrolithiasis accompanied frequently this anomaly. Calculi in dystopic kidneys accounted for 39.8 per cent of his cases of nephrolithiasis in various types of anomalous kidneys. Ostropolskaya (1970) reported 15 children with hydronephrosis of dystopic kidneys.

It is our experience that dystopic kidneys are often involved in various diseases. We encountered this in 82.4 per cent of our patients, whereas in the remaining 17.6 per cent renal dystopia was asymptomatic.

We observed various diseases in dystopic kidneys. Most of them were ailments associated with urinary stasis in the pelves (hydronephrosis, nephrolithiasis and pyelonephritis), and were encountered in 45 per cent of patients. Hydronephrosis accounted for 50 per cent of the diseases, nephrolithiasis for 21 per cent, and pyelonephritis as an independent disease for 20.3 per cent. We also observed hypernephroma in 2.9 per cent of cases and nephroptosis in 5.8 per cent. In 28.4 per cent of our patients the dystopic kidney caused pain; thorough examination of these patients failed to reveal any disease. In these cases the dystopic kidney itself without any marked disease was the cause of the pain. It is quite probable that these patients had urinary stasis in the pelves of the dystopic kidneys, but the disease (hydronephrosis) had not yet become evident. Urokymography and cineurography showed that the pain was due to dyskinetic disturbances in the upper urinary tract.

Thus, dystopic kidneys are predisposed to the development of retention diseases. Other diseases are apparently encountered no more frequently than in normally developed kidneys.

Symptomatology, clinical course and recognition of dystopic kidneys. A dystopic kidney uncomplicated by disease may produce clinical symptoms. This was noted in 28 per cent of our patients.

The symptoms of dystopic kidney are varied and vague. They are often atypical and mild, particularly in lumbar dystopia, which is why the anomaly remains unrecognized in many cases.

The lower the position of the dystopic kidney, the more evident are the symptoms. The different signs are caused by the direct mechanical pressure of the malposed kidney on the adjacent organs, vessels, nerve ganglia and plexuses, and the reflex effect on the function of different organs through the nerve plexuses. At the same time one cannot ignore the dyskinetic disorders of the upper urinary tract and distention of the pelvis observed in these cases. All these causes may explain the disorders of the nervous system and female genitals, and dysuric and dyspeptic signs encountered in renal dystopia. Another symptom is a tumour palpated in the abdomen.

Pain is the main and often the only symptom in patients with dystopic kidneys. We encountered it in 59 per cent of patients with the anomaly uncomplicated by any severe diseases. In 39 per cent of cases dystopic kidneys were asymptomatic and were revealed by pure chance during examination.

Pain was felt during movement, in walking, and on physical exertion, and abated when the patient was at rest. In some cases patients relieved it by lying on their stomach.

The pain was dull and constant or sharp and occurring in attacks. Sometimes patients noted only a feeling of heaviness in the lower abdomen. The renal colic syndrome is relatively rare and is most frequently encountered in lumbar dystopia and may be associated with circulatory disorders in the kidney.

The localization of the pain is extremely variable. Pain in the small of the back occurs in lumbar dystopia and very rarely in iliac dystopia. In iliac and pelvic dystopic kidneys pain is usually experienced in the abdomen, in its lower parts, in the region of the sacrum and pelvis. In some cases it is referred from there to the lower limbs.

Such indefinitely localized and atypical pain differs sharply from the commonly encountered renal pain. It resembles the pain usually observed in appendicitis, ileus, diseases of the intestine and the female genitals; a pain localized in the thorax resembles the pain felt in diseases of the heart and lungs. The following cases from our practice are given in illustration.

Ts., a 34-year-old woman, was admitted to the clinic on May 7, 1956. She complained of a dull, constant pain in the left iliac region from which she had been suffering for a month. From time to time she had nausea and constipation. A smooth, elastic and painless tumour was palpated in the left iliac region. An earlier examination of the gastro-intestinal tract had failed to reveal any abnormality. Blood and urine tests showed normal values. Excretory urography demonstrated a normal pyelo-calyceal system on the right; but the left pelvis was not found at its normal site. The shadow of the ureter was clearly visible in the minor pelvis on the left and terminated in the shadow of the pelvic bones. Retrograde pyelography demonstrated an anomalous pelvis in the left iliac region with calyces directed at the ver-

tebrae. The ureter of this kidney was shortened. No signs of renal disease were revealed. The diagnosis of iliac dystopia of the left kidney was made.

The unusual localization of pain in the left iliac region in this patient and the dyspeptic disorders and constipation suggested a disease of the gastro-intestinal tract. The tumour revealed in this region provided grounds for suspecting a dystopic kidney and undertaking a urological examination.

B., a 27-year-old woman, was transferred to the clinic on October 18, 1967 from the gynaecological department. She had been subjected to laparotomy because a tumour of the uterine appendages was suspected. A tumour was found in the retroperitoneal space of the pelvis during the operation and it was presumed that the patient had a dystopic kidney. She had no complaints except for a dull pain felt now and then in the lower abdomen. On examination the gynaecologist had palpated a tumour in the pelvis, which led to the erroneous diagnosis of a tumour of the genitals and to an unjustified operation. Blood and urine tests showed normal values. Excretory urography showed the right kidney to be in a normal position, with a normal pyelo-calyceal system. The shadow of the pelvis of the left kidney was seen on the left as an irregularly shaped spot against the background of the sacrum. Retrograde pyelography showed the pelvis of the left kidney on the left against the background of the sacrum; its calyces were directed upward, its ureter was short. No other abnormalities were revealed in the kidney. A diagnosis of pelvic dystopia of the left kidney was made.

In this case there was a mild dull pain in the lower abdomen which brought the patient to a gynaecologist for examination. Due to the presence of a tumour in the pelvis a new growth of the genitals was suspected and an unjustified operation performed. With timely urological examination this operation could have been avoided.

In crossed renal dystopia the pain may be localized on the contralateral side. This sometimes causes difficulties in diagnosing the condition.

In cases of renal dystopia a **tumour** is often palpated in the abdomen through the vagina or rectum in a low position or above the pubis or in the iliac and lumbar regions in a higher position. A dystopic kidney is felt as an oval tumour, which is tender to pressure and poorly mobile. It usually has pulsating vessels on its surface. This tumour lies close to the midline as a rule.

Dysuric signs in renal dystopia are commonly associated with inflammatory changes in the urinary tract. In pelvic dystopia, however, there may be frequent micturition with no abnormalities in the urinary tract. Gimpelson (1949) attributes this symptom to compression of the urinary bladder by the dystopic kidney.

Disorders of the female genitals evident in disturbances of the menstrual cycle may also be explained, in Gimpelson's (1949) opinion, by pressure of the dystopic kidney on the uterus. Neuro-

logical disorders may also be of significance here. We found this symptom in several of our patients.

Dyspeptic disorders, which are a frequent occurrence in patients with renal dystopia, could be linked in many instances with gastro-intestinal diseases, but thorough examination fails to reveal these diseases in some patients with the anomaly. These patients complain of persistent constipation and bouts of spastic pain in the region of the stomach or intestine. Punin (1946) claims that this is a consequence of the pressure of the dystopic kidney on the adjacent intestine and its vessels and nerves. The close reflex connection between the kidneys and the gastro-intestinal tract due to the large number of anastomoses between the renal and mesenteric nerve plexuses is evidently of importance here. Stimulation of these plexuses by the malposed kidney may cause dyspeptic symptoms and simulate gastro-intestinal diseases.

Dyspeptic disorders are sometimes the only symptom of a dystopic kidney. In our cases they were mistaken for signs of gastro-intestinal disease (appendicitis, ileus, colitis, etc.). They create considerable difficulties in differential diagnosis.

Disorders of the nervous and cardiovascular systems are frequently encountered in patients with dystopic kidneys. Most of them are evidently linked with vegetative dystonia due to reflex influences from nerve plexuses against which the anomalous kidney presses. Vegetative dystonia may explain many symptoms such as abdominal pain, dyspeptic disorders, attacks of palpitation, etc. Punin (1946) contends that these disorders may take on a psychotic character.

Some patients with pelvic renal dystopia have varicose veins of the lower limb on the side of the anomaly; paraesthesia of the lower limb and increased venous pressure are sometimes encountered.

Profuse haematuria sometimes occurs in dystopic kidneys in which no diseases have been revealed despite thorough examination.

G., a 32-year-old woman, was admitted to the clinic on February 14, 1965 complaining of blood in the urine. The blood had appeared suddenly. No other symptoms were revealed. All urine and blood tests showed normal findings with the exception of a single urinalysis which revealed a large number of erythrocytes. A complete urological examination was undertaken, which helped make a diagnosis of iliac dystopia of the right kidney. No urological diseases were found. A follow-up of seven years also showed no disease in the dystopic kidney.

The appearance of blood in the urine of this patient with renal dystopia may be attributed to a temporary disturbance of the passage of urine in the pelvis and fornical haemorrhage. The weakly protected dystopic kidney situated above the sacroiliac articulation is more vulnerable to injury. It is possible that the

profuse haematuria in these cases is caused by a mild trauma of the kidney which was unnoticed by the patient.

Clinical course. According to the clinical course two groups of patients with renal dystopia can be distinguished: (1) a group without diseases and pathological disorders, and (2) a group without diseases but with pathological disorders.

Among our patients 1.76 per cent had asymptomatic renal dystopia. According to Gimpelson (1949), in 8 of 51 patients the dystopic kidney was found accidentally during prophylactic surveys or urological examination for diseases of the other kidney or lower urinary tract.

The subsequent follow-ups of patients of this group showed no pathological changes in the kidneys. But a follow-up period of over ten years showed that some patients of this group developed diseases of the dystopic kidney. Some of them underwent an unjustified operation because the anomalous kidney was mistaken for a tumour of the abdominal cavity or the female genitals. After surgery some of these patients had dull pain which could be attributed to the anomalous kidney. These patients were transferred to the group without disease, but with pathological disorders.

The symptoms of dystopic kidneys listed above without disease were encountered in 28.4 per cent of our clinical cases. Gimpelson (1949) observed them in nine of his 51 patients. These symptoms develop in the patient in childhood or later in adolescence, or in old age. In our experience, however, this condition was revealed predominantly in persons of middle age, when the organism functions with the greatest effort. It is quite possible that this is due to the weakening of the organism by disease, as was the case with some of our patients.

In such cases renal dystopia takes a chronic clinical course with mild symptoms. The following case is given in illustration.

Z., a 27-year-old woman, was admitted to the clinic on March 23, 1957. She complained of dull pain in the right half of the abdomen which appeared during physical exertion. She had been ill since childhood with the dull abdominal pain sometimes giving way to sharp attacks. She had also suffered from nausea, constipation and episodes of palpitation. At the age of twelve she underwent appendectomy which, however, brought no relief. She was treated for various gastro-intestinal diseases, but with no effect. At one of her examinations a tumour was palpated in the right iliac region and nephroptosis was suspected. With this tentative diagnosis the patient was admitted for urological examination. Blood and urinary tests showed normal values. X-ray urological examination disclosed a dystopic kidney in the right iliac region; its calyces were directed at the vertebrae, the pelvis was positioned laterally and the ureter was short. No signs of disease were in evidence. The patient has been kept under our surveillance for 17 years: the complaints mentioned above appear once or twice a year, the kidney functions well, the pelvis is somewhat distended.

This case is characterized by symptoms of renal dystopia persisting for many years. The mild manifestations of the anomaly hardly trouble the patient and there is therefore no need for surgery. In some patients dyskinesia of the upper urinary tract and urinary stasis in the pelves develop with time, and the disease calls for special treatment.

One opinion appearing in the literature is that a dystopic kidney in the pelvis compresses the uterus and hinders delivery of an infant (Sokolov, 1928). Feigel (1926) describes rupture of the uterus during delivery in a woman with renal dystopia and, on this basis, recommends caesarean section or the induction of premature labour for these parturients and even nephrectomy at an early stage of the pregnancy. Shakhbazyan (1931) claimed that caesarean section was better than operation on the kidney during pregnancy. In contrast, Thompson and Pace (1937) and Anderson et al. (1951) claimed that women with dystopic kidneys had no particular complications during pregnancy and delivery. It is our experience that in women with pelvic kidney dystopia, pregnancy and delivery usually follow a normal course if there are no additional anomalies of the pelvic bones. If a first delivery is normal, subsequent deliveries will also be favourable. Some of our patients with kidney pelvic dystopia gave birth to three normal children. That is why we do not feel free to recommend abortion or surgical operations in cases of renal pelvic dystopia. Indications for such procedures may occur only in diseases of the anomalous kidney.

DIAGNOSIS OF KIDNEY DYSTOPIA

The symptoms of the dystopic kidney discussed above are atypical of a pathological renal condition and the diagnosis of such a renal anomaly is therefore often delayed. Gimpelson (1949) claims that dystopia of the kidney is suggested by such varied symptoms.

The medical history helps in the diagnosis of very rare cases when patients had been examined or operated on earlier and know that they have an anomalous kidney. There were only two patients aware of their condition in our practice. Laboratory and instrumental methods of examination yield no characteristic data as a rule, although cystoscopy may sometimes show an asymmetric urinary bladder in a person with pelvic dystopia of the kidney.

Palpation gives rise to the suspicion that the patient has this renal anomaly, but in lumbar and thoracic dystopia the kidney

cannot usually be palpated and such an examination is therefore devoid of significance in these cases. This is why anomalies of this localization are often discovered by chance during other examinations or operation. In pelvic and iliac positions the kidney can often be palpated. A tumour, resembling a kidney in size, felt in the pelvis close to the midline should cause one to consider the possibility of renal dystopia. Israel was the first in 1901 to make a diagnosis of a dystopic kidney on the basis of this sign; laparotomy verified the diagnosis.

Abdominal pain of indefinite localization or pain in the lower abdomen, in some cases dysuric disorders, various symptoms of the female genitals and the gastro-intestinal, nervous and cardiovascular system and, in addition, a tumour palpated in the abdomen should arouse the suspicions of the physician and call for urological examination to rule out renal dystopia, especially before laparotomy. This applies, in particular, to cases in which the disease has followed a chronic course over many years.

Other methods of examination such as inspection for pulsation on the anterior surface of the kidney and palpatory proteinuria are unreliable and even detrimental for the patient. Pulsation of the vessels may also be detected in a case of a retroperitoneal tumour. Palpatory proteinuria causes excess traumatization of a kidney even if it is healthy. With the development of X-ray methods of examination these manipulations have lost their clinical significance.

X-ray methods of examination are the leading techniques in the diagnosis of dystopic kidneys.

Scout roentgenoscopy and roentgenography may suggest a diagnosis of renal dystopia when the outlines of one of the kidneys are not seen on the radiograph in the lumbar region but are traced at the midline of the body at the level of the distal lumbar vertebrae. In some cases the radiographs show the liver to be in a low and vertical position, in which case it is evidently occupying the space vacated by the kidney.

In thoracic renal dystopia radiographs show a shadow above the diaphragm against the background of the pulmonary shadow. Anteroposterior and lateral view radiographs show an oval tumour in the dorsal region; the tumour is homogeneous and incompletely separated from the diaphragm during respiration. The surrounding pulmonary tissue is normal.

It is difficult to decide on the possibility of the patient having a dystopic kidney on the basis of the fact that a catheter can be introduced into the kidney for only a short distance, because it may encounter an obstacle in the ureter. The X-ray technique with a metal ring fitted around the palpated tumour is often

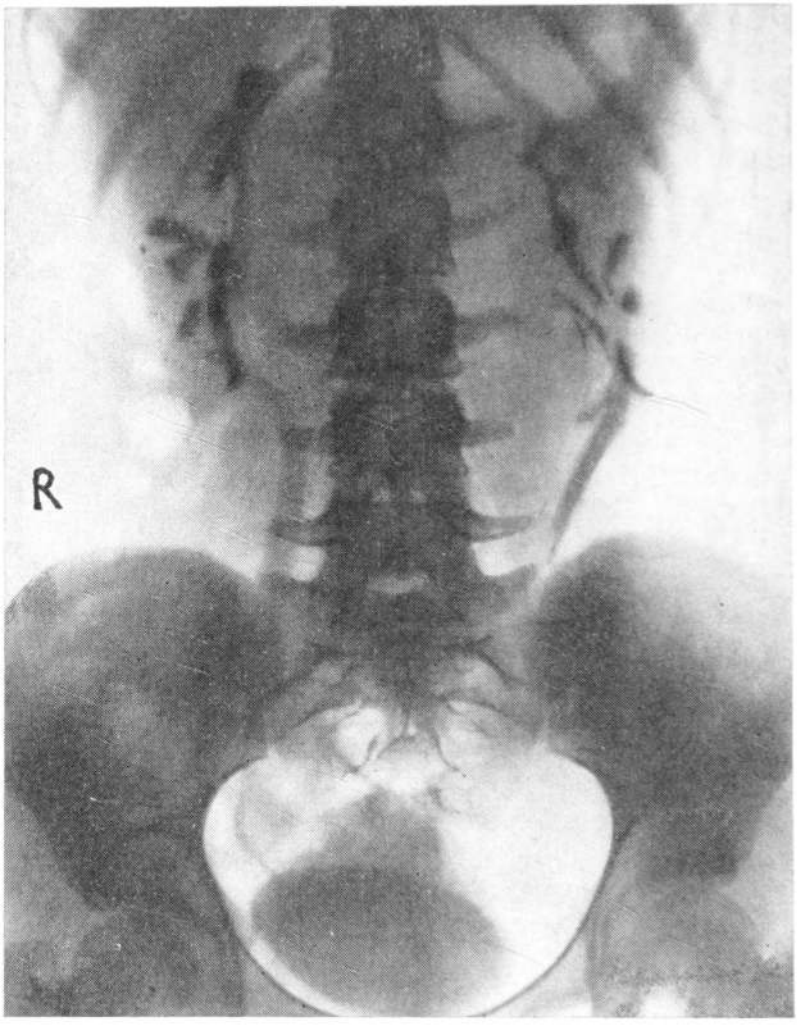


Fig. 72. Lumbar kidney dystopia. Infusion urography. The calyces of the dystopic kidneys are located medially in relation to the renal pelvis

unreliable, because the ring may only embrace the pole of the kidney while the rest of the kidney is not palpable.

In lumbar renal dystopia a catheter introduced into the ureter is sometimes displaced medially in the proximal part. This may be interpreted as indication of a median position of the pelvis and suggests lumbar dystopia of the kidney.

Excretory urography, the infusion technique in particular, makes it possible to confirm or rule out the diagnosis when the kidney is functioning normally. The diagnosis of the anomaly, i.e. renal dystopia, can be made with confidence on the grounds of the lateral position of the pelvis, median position of the calyces and a short ureter (Fig. 72).

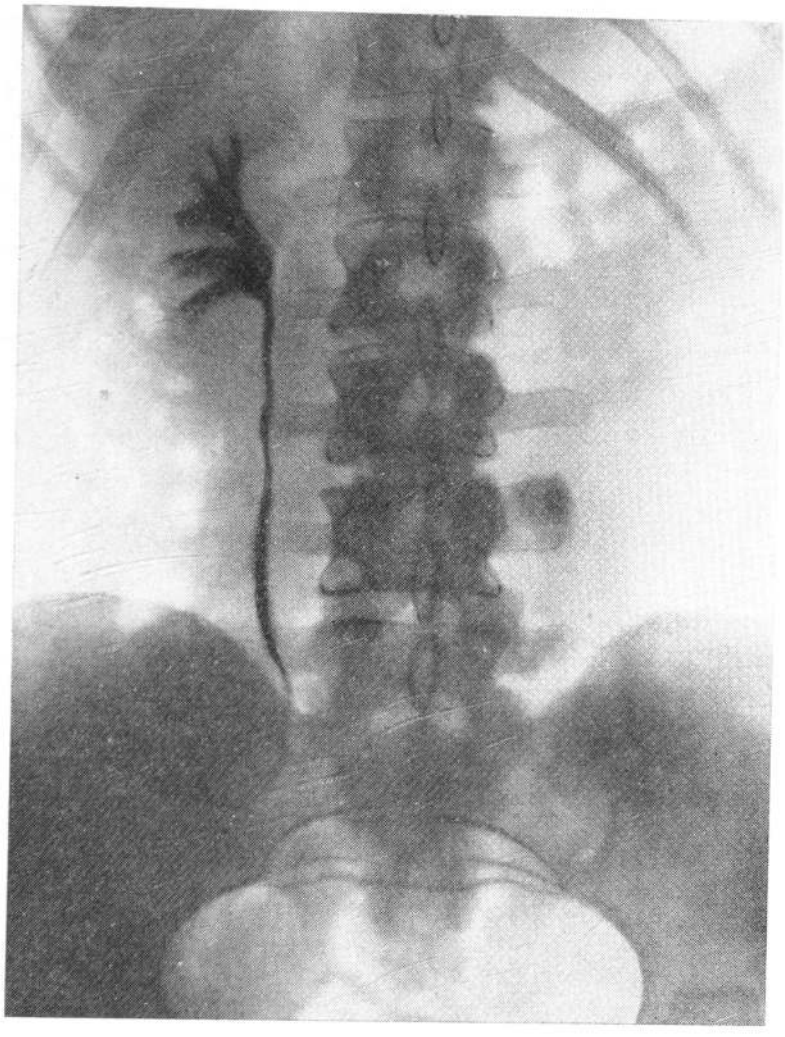


Fig. 73. Case S. Lumbar kidney dystopia. Excretory urography. The shadow of the upper calyx of the dystopic kidney is seen at the transverse process of the fourth lumbar vertebra on the left

Even when the dystopic kidney functions well, the shadow of its pelvis is often not shown in excretory urography. This occurs because the upper urinary tract is above the bones of the pelvis and the shadows of the renal pelvis and calyces cannot be seen. In such cases the functioning kidney can be judged by filling the ureter with a contrast medium, the image of the pelvis being sought by other means. Sometimes it proves possible to see the shadaw of a calyx against the bones of the pelvis or vertebrae (Fig. 73). Excretory urography may reveal a dystopic renal pelvis in the thorax and thus help to make a diagnosis.

Thus, even the infusion technique fails in some cases to produce an image of the upper urinary tract of a kidney in cases of

thoracic or lumbar dystopia and particularly in iliac and pelvic dystopia. Such cases call for retrograde pyelography.

Retrograde pyelography helps to make a proper diagnosis of a dystopic kidney from the image of the upper urinary tract. With the wide use of this method renal dystopia is no longer a rare finding.

Retrograde pyelography produces a clearer image of the urinary tract and, consequently, provides more convincing data for a diagnosis of the anomaly and secondary changes in the kidney. It helps in differentiating deformity and distension of the calyces due to renal anomaly from those due to diseases of the kidney.

According to Gimpelson (1949), the pyelogram of a dystopic kidney shows the low position of the kidney, an unusually shaped renal pelvis, the medial position of the calyces, and the lateral or middle origin of the ureter and its shortening. The pelves of the dystopic kidney often lie close to the midline of the body. This is particularly obvious in pelvic and iliac renal dystopia. The pelves and calyces of dystopic kidneys vary considerably in each case, a fact illustrated in Figs. 74-80.

To make a correct diagnosis of crossed and bilateral dystopia it is necessary to perform bilateral retrograde pyelography, which is undertaken in two stages because complications may develop. This excludes any diagnostic error.

When in excretory urography both pelves cannot be clearly seen on the radiographs, the diagnosis cannot be made without resorting to bilateral retrograde pyelography.

P., was admitted to the urological clinic on March 15, 1966, complaining of dull abdominal pain. Two elastic, smooth and mobile tumours were palpated below the umbilicus. He said that in the past few years he had experienced sharp pain in the right abdomen and right lumbar region after physical exertion. Between these attacks he had had dull abdominal pains. Excretory urography showed traces of a shadow in the form of an irregularly shaped spot on the right above the iliac wing. On the left the upper urinary tract could not be seen. Chromocystoscopy revealed no abnormalities and indicated good functioning of both kidneys. Bilateral retrograde pyelography revealed bilateral renal dystopia, lumbar on the right and iliac on the left. Pyeloscopy showed restricted mobility of the right kidney. The kidneys are apart from each other. Nephropexy was performed on the right side. The patient recovered.

Bilateral retrograde pyelography helped us in this case to arrive at the proper diagnosis of bilateral renal dystopia and rule out a disease of the kidney. Fusion of the kidneys was ruled out by pyeloscopy and during subsequent nephropexy.

Retrograde pyelography should therefore be used in renal dystopia when the findings of excretory urography are doubtful and also when it is necessary to gain detailed information on

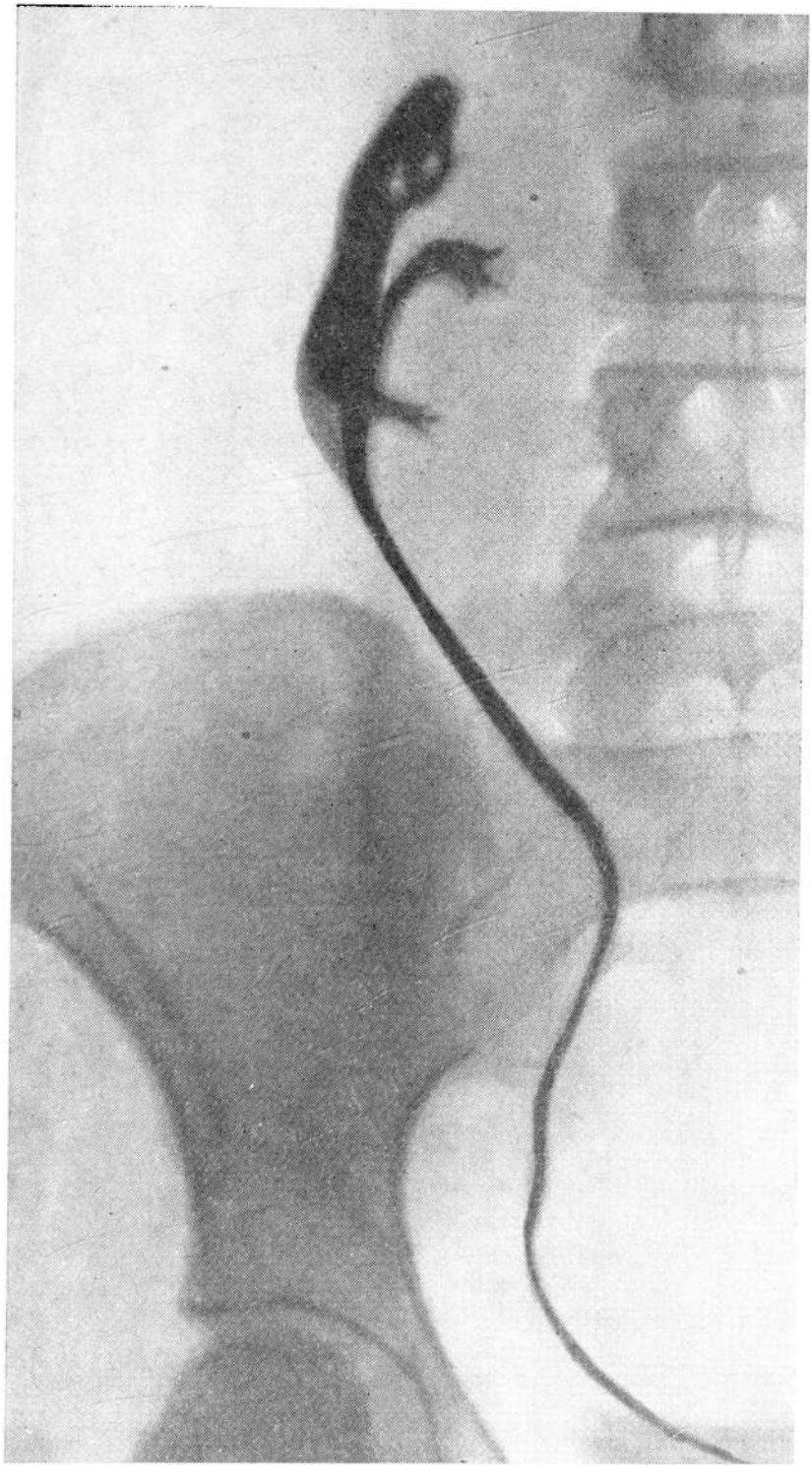


Fig. 74. Case M. Lumbar dystopia of the kidney on the right. Retrograde pyelography

Fig. 75. Case M. Lumbo-iliac dystopia of the kidney. Retrograde pyelography

Fig. 76. Case L. Iliac dystopia of the kidney. Retrograde pyelography

the structure of the urinary tract and rule out or confirm an ailment of the dystopic kidney before surgery. Retrograde pyelography is also applied when the results of excretory urography are questionable and bilateral renal dystopia is to be diagnosed. Bearing in mind the hazards of retrograde pyelography, and of bilateral ureteral catheterization in particular, one should try to make the diagnosis by means of excretory urography and apply retrograde pyelography as a last resort.

Diagnostic pneumoretroperitoneum yields information on the size, shape and position of the dystopic kidney. Our research

Fig. 77. Case M. Pelvic dystopia of the kidney. Retrograde pyelography

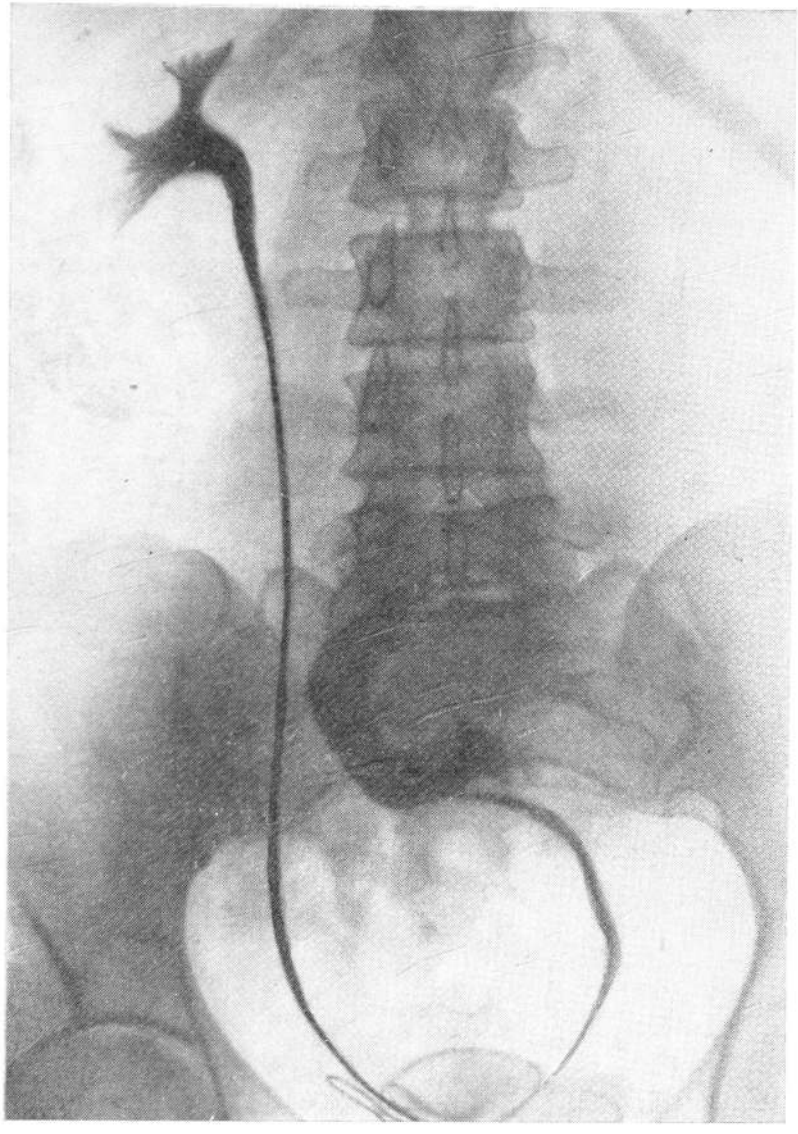


Fig. 78. Case P. Crossed dystopia of the left kidney. Retrograde pyelography

(Shimkus, 1961) showed that pneumoretroperitoneum makes it possible to appraise the condition of the renal parenchyma and the position of the kidney in relation to other organs and tissues before surgery. When excretory urography is ineffective and retrograde pyelography cannot be performed because of an obstacle in the ureter, retroperitoneal introduction of gas helps to reveal the image of the kidney and provides an idea of the amount of preserved parenchyma.

A renal anomaly can be assumed from the shape of the kidney. Pneumoretroperitoneum also helps in differentiating fused kid-

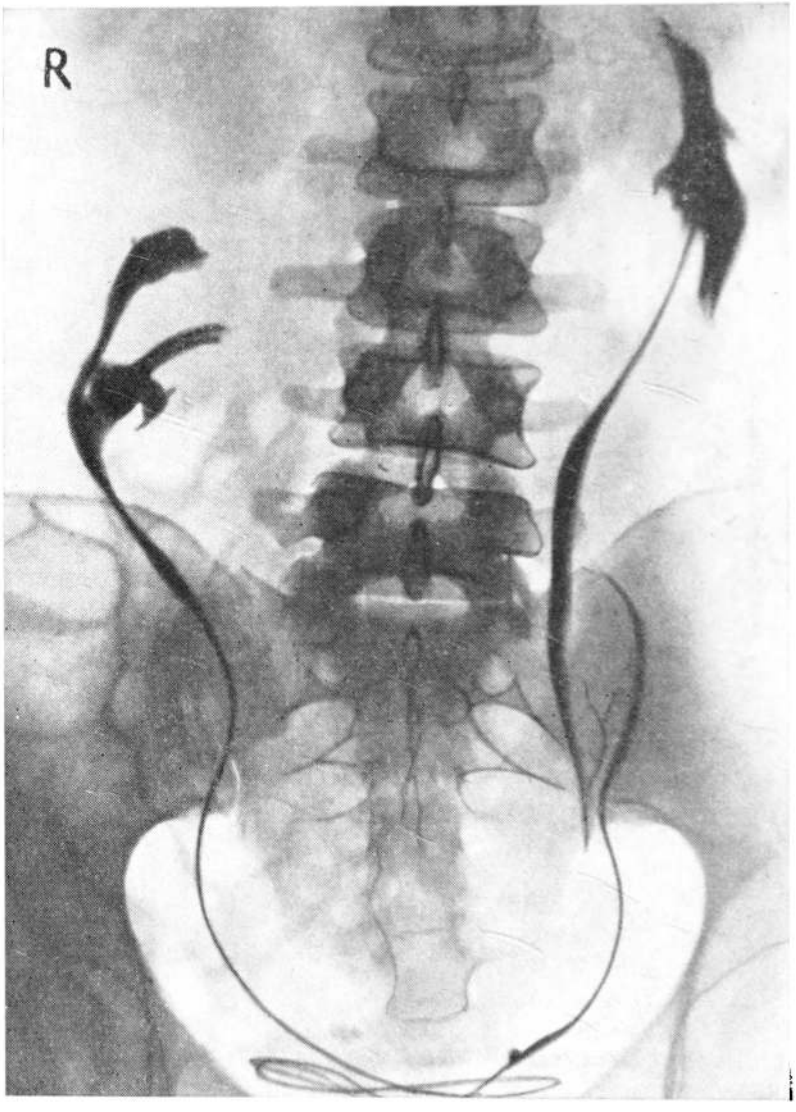


Fig. 79. Case M. Bilateral lumbar dystopia of the kidneys. Retrograde pyelography

neys from bilateral renal dystopia with no fusion of the kidneys. This is particularly effective when radiographs are taken at an angle of 45 degrees. Pneumoretroperitoneum reveals signs of sclerotic paranephritis. In such cases there is no image of the kidney on the radiographs. According to Franciscovic and Martincic (1959), in thoracic renal dystopia pneumoretroperitoneum shows the shadow of the anomalous kidney above the diaphragm.

In lumbar renal dystopia the lobate nature of the kidney is clearly seen on the radiographs (Fig. 81). In iliac dystopia the shape of the kidney is also well discerned, but the lobate charac-

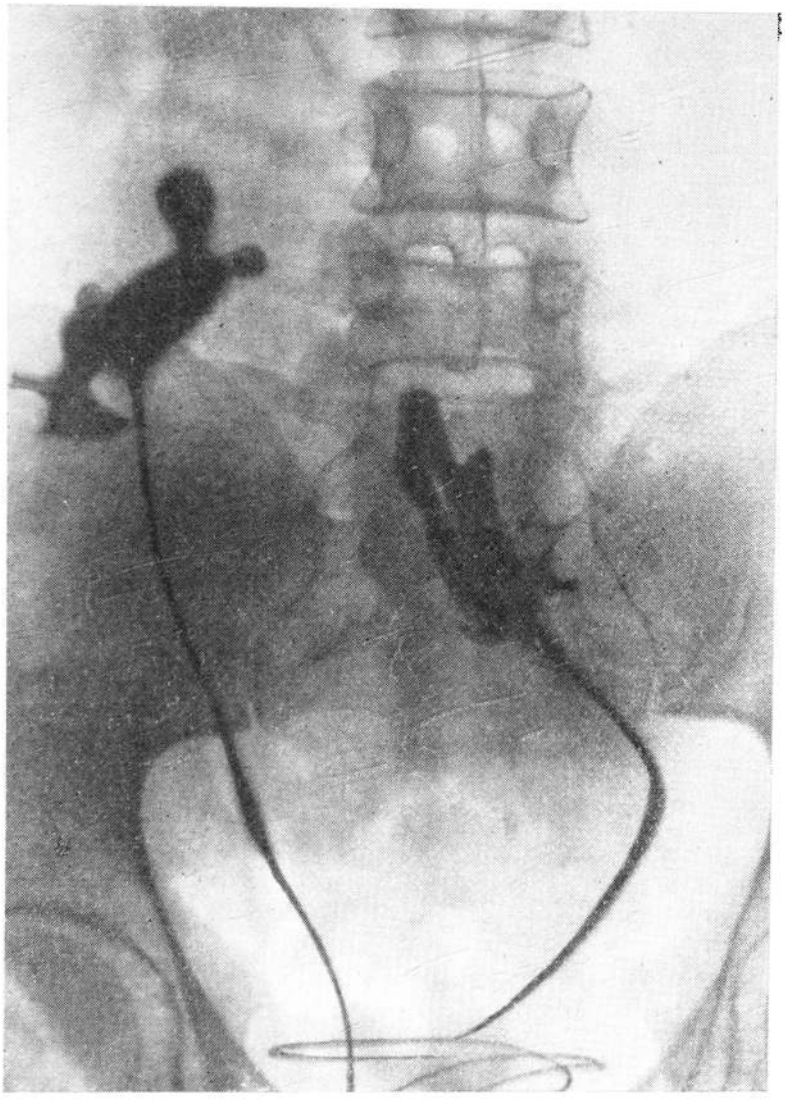


Fig. 80. Case P. Bilateral renal dystopia, lumbar on the right and iliac on the left. Retrograde pyelography

ter of the organ is less evident. The position of the kidney below the normal level and its somewhat enlarged dimensions are the most common signs of dystopic kidneys in these radiographs.

Radiographs produced by a combination of pneumoretroperitoneum with excretory urography are particularly demonstrative. They show the size of the kidney, its position, relation to the adjacent organs, size and function. With the use of the infusion technique of excretory urography the information gained by means of these two methods often makes it possible to avoid the use of retrograde pyelography.

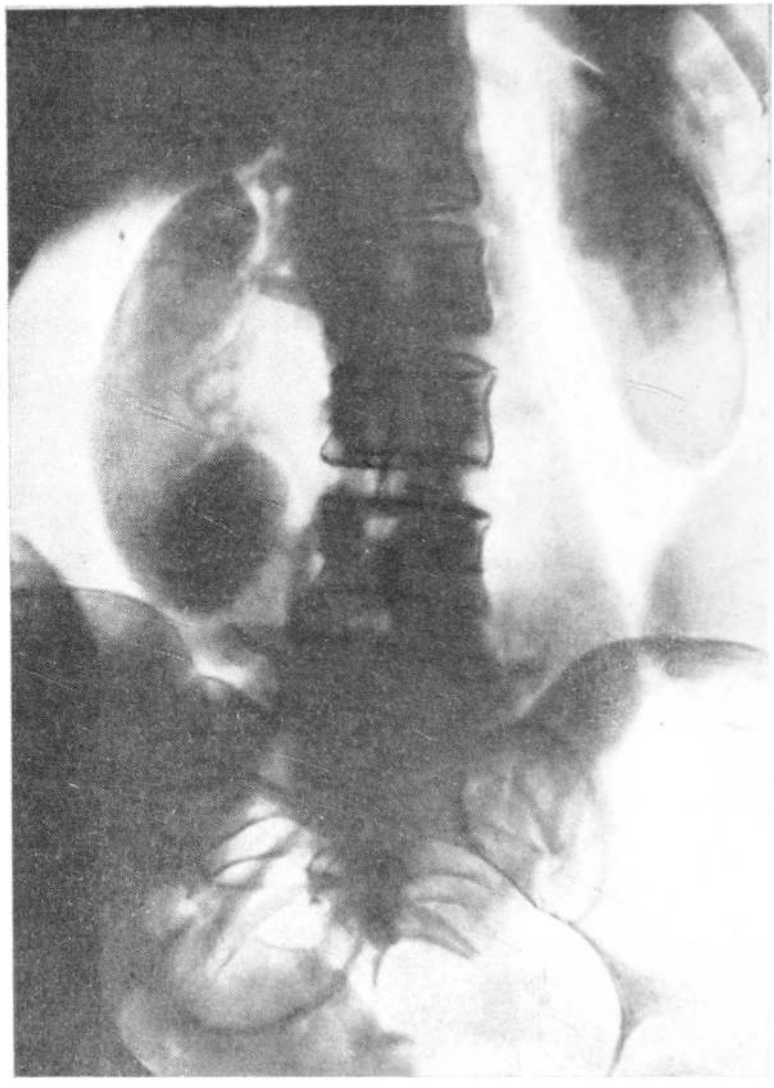


Fig. 81. Case D. Lumbar dystopia of the kidney. Pneumoretroperitoneum. Lobate character of the kidney

Pneumoretroperitoneum performed in combination with retrograde pyelography produces the most demonstrative radiographs (Fig. 82). They give a good image of the kidney and its urinary tract and the thickness of the preserved renal parenchyma.

The combined use of pneumoretroperitoneum with tomography helps in the diagnosis of the different forms of renal dystopia. In renal anomalies tomography should be made in the second of three planes from the table. Pneumoretroperitoneum gives a clear image of the organ while tomography determines its exact position and excludes the possibility of deficient examination associated with poor preparation of the patient. On these

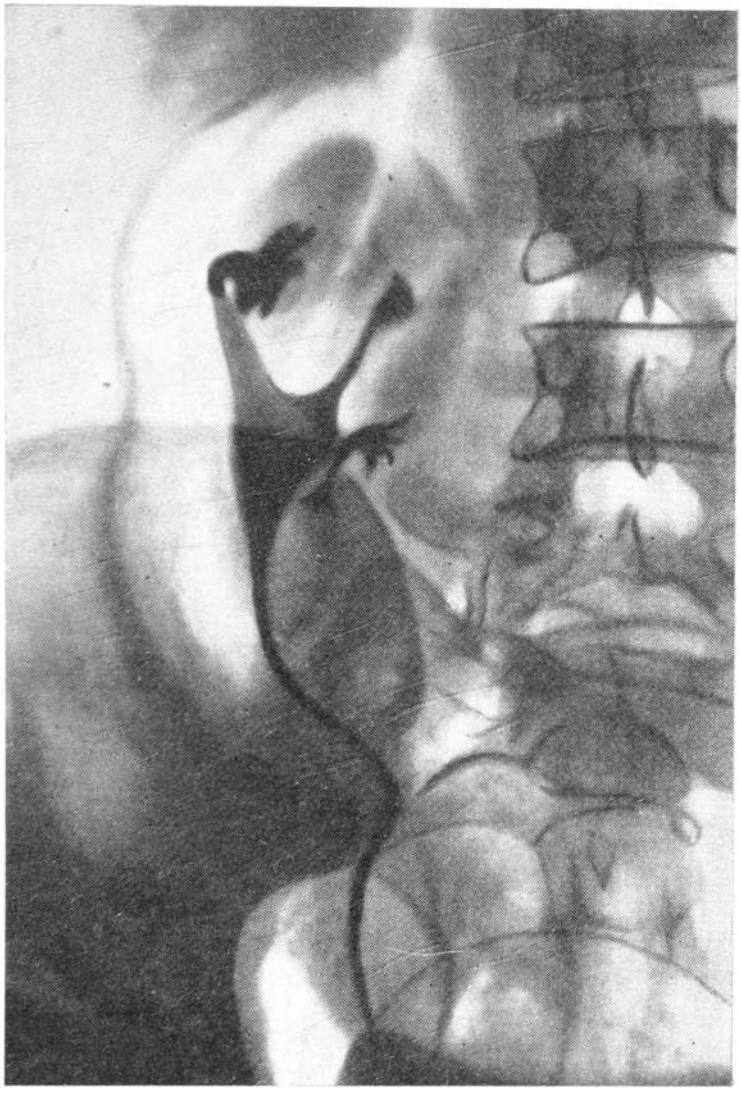


Fig. 82. Case Z. Iliac dystopia of the kidney. Pneumoretroperitoneum and retrograde pyelography

radiographs the dystopic kidney is characterized by a low position and closeness to the vertebrae.

From our experience we recommend that these methods of examination be used in the diagnosis of dystopic kidneys and particularly for ruling out or confirming fusion of anomalous kidneys.

Renal angiography in dystopic kidneys yields valuable information on the kidney's function, shape and position and the character of its blood supply. Lopatkin (1961) presented convincing data on the diagnostic possibilities of this method in cases

of kidney anomalies. Szporny et al. (1971) point to the value of renal angiography in pelvic dystopia of the kidneys.

Renal angiography is called for in determining the type of kidney malposition when this cannot be ascertained by excretory urography, and in determining the character of the blood circulation in it the knowledge of which is essential before the operation. This examination may provide more accurate information on the length of the renal vessels. If they are short nephropexy will not be undertaken because under these circumstances the operation will not only fail to bring relief, but may prove hazardous for the patient. The method helps to differentiate a dystopic kidney from a downward displaced kidney.

Renal angiography applied in combination with pneumoretroperitoneum far exceeds the possibilities of routine examination techniques. The two methods should be combined when the findings of tomography and pneumoretroperitoneum give rise to doubt.

The results of renal angiography in dystopic kidneys reveal the difficulties which can be expected during the operation, determine which vessels are to be ligated, and help to avoid undue haemorrhage.

Pyeloscopy, urokymography and cineurography are particularly important in the diagnosis of dystopic kidneys, because the anomaly often causes dyskinesia of the upper urinary tract. These methods of examination also help to differentiate fused kidneys from bilateral renal dystopia by disclosing or ruling out conjunction of the organs, since kidneys which are not joined to each other move apart.

On radiographs taken by means of urokymography we often noted marked lateral displacements of the upper third of the ureters and weakened contraction of these segments. Quite often the contractions of the pelvis of the anomalous kidney are also weak. These changes are evidently caused by compression of the prepelvic segment of the ureter above the lower pole of the dystopic kidney. It is possible that it is precisely these dyskinetic disturbances which are the cause of the pathological disorders induced by what seemed to be a healthy dystopic kidney. We consider this condition to be the initial manifestation of the disease. Later, urinary stasis in the pelves and hydronephrosis develop in these patients, which is why it is necessary to begin treatment of the disease at this stage. Cineurography and urokymography enable us to confirm changes in the urodynamics of the upper urinary tract of patients with dystopic kidneys

Isotope renography and kidney scanning are important methods of diagnosis of dystopic kidneys. Isotope renography gives an idea of the function of the kidney and in some cases indicates

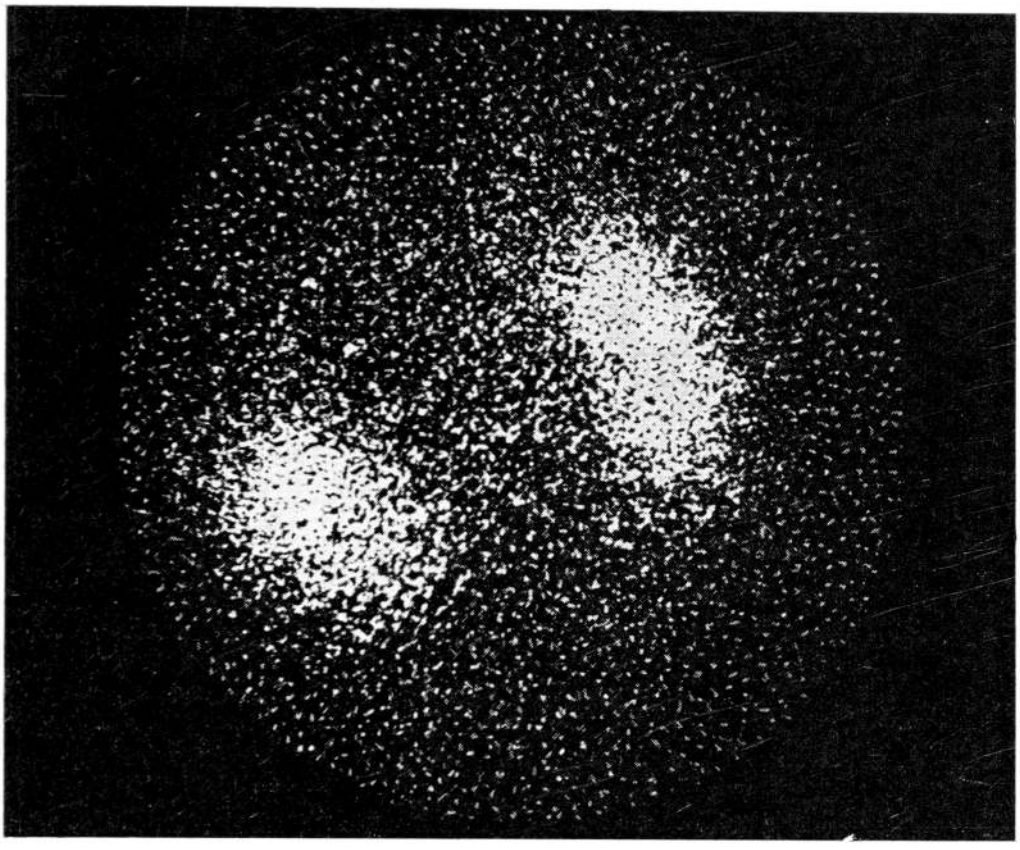


Fig. 83. Dynamic scintigraphy. Lumbar kidney dystopia

the absence of function (if the sensors are placed on the site usually occupied by the normal kidney) although the function in fact is quite satisfactory. This fact alone forces the physician to carry out an urological examination and so discover the anomalous kidney. If the existence of the dystopic kidney is known prior to examination, correct positioning of the sensors gives an idea of the function of the kidney.

The position, size and function of the kidney can also be determined by scanning (Figs. 83-84). It is our experience that this method is valuable in the recognition of dystopic kidneys, the more so since in using other methods of examination dystopic kidneys have to be differentiated from downward displaced kidneys.

The use of this method is particularly interesting in cases of crossed dystopia of the kidneys. Then the outlines of both kidneys are seen to one side of the midline. Lurie et al. (1971) verified crossed kidney dystopia in one of their cases by scanning.

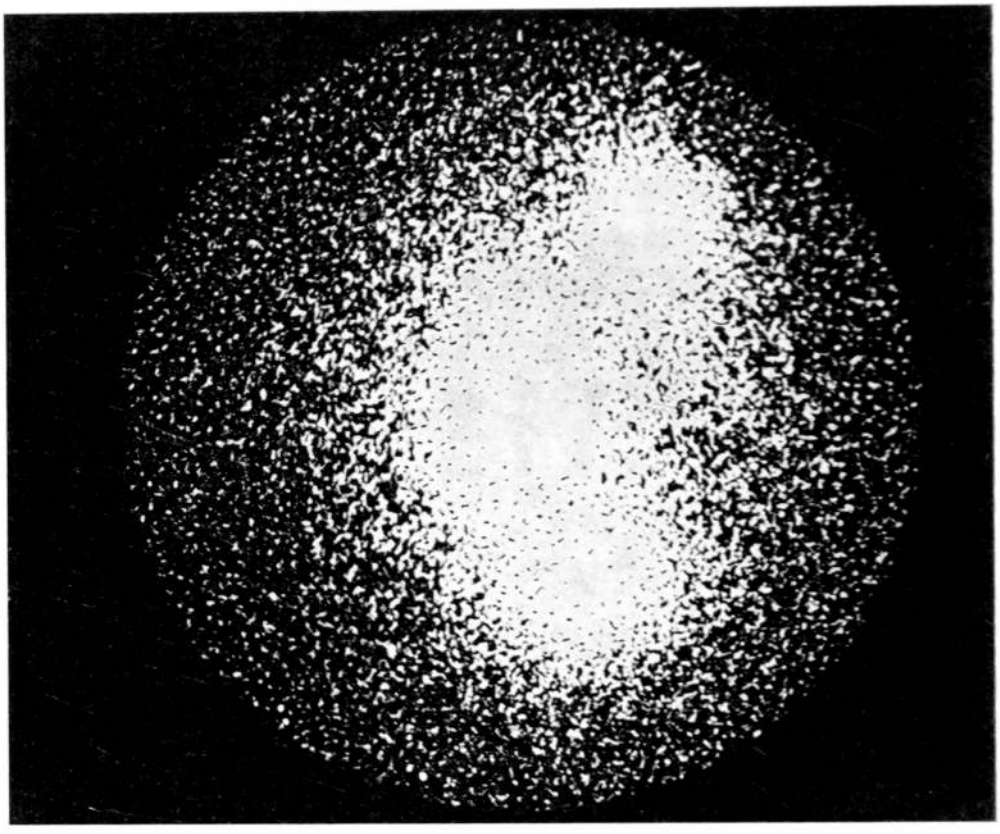


Fig. 84. Dynamic scintigraphy. Pelvic kidney dystopia

Our experience shows that a healthy dystopic kidney may be differentiated from diseases of an anomalous kidney by scanning. That is why we believe it necessary to use this method of examination in every case of dystopic kidneys. In cases of bilateral dystopia it might suggest the possibility of fused kidneys.

Planning for the examination of a patient who is suspected of having a dystopic kidney should include a progressive use of methods ranging from the simple to more complex so as to make a diagnosis and rule out any disease with the least possible discomfort for the patient. When the results of objective methods of examination give reason to suspect the anomaly, we make a scout radiograph of the lumbar region and the lungs. A malposed outline of the kidney may sometimes be seen on it. After that excretory urography is performed. In most cases it helps to identify high kidney dystopia. In iliac and pelvic dystopias it is necessary to resort to retrograde pyelography. Renography and scanning of the kidneys provide information on the function of the organ, whereas cineurography yields data on the function of the upper urinary tract. Telepyeloscopy is carried out when

it is necessary to differentiate fused kidney from bilateral renal dystopia. When the results of the examination cause doubt, tomourography or diagnostic pneumoretroperitoneum is performed. When the results of all these methods are questionable, renal angiography should be resorted to. This method is also used with some patients before operating because it provides valuable information for the surgeon.

Diagnostic Errors and Differential Diagnosis

Dystopic kidneys often lead to errors in diagnosis. No other organ is so subjected to unwarranted operations as the anomalous kidney. The literature includes reports of a dystopic kidney being mistaken for a retroperitoneal tumour, tumour of the minor pelvis or mesentery, enlarged lymph nodes, ovarian cyst, tumour of the uterus or its appendages, mesadenitis, appendicular infiltrate, appendicitis, ileus, gastric or duodenal ulcer, duodenal diverticulum and aneurysm of the aorta. Since errors are being made continually, the problem should be brought to the attention of a wide circle of surgeons and gynaecologists.

With the development of thoracic surgery there are more and more reports in the literature on unwarranted thoracotomy in cases of thoracic renal dystopia. Most of the described renal anomalies have been recognized during such operations. Quite frequently we learn of the removal of a solitary dystopic kidney. Andersen and Harrison (1965), for instance, reported two cases of unintentional nephrectomy of a solitary dystopic kidney. Subsequent treatment by means of kidney transplantation proved ineffective.

Among the various renal anomalies, dystopic kidneys cause diagnostic errors most frequently. Because of their low position they are accessible to palpation and often cause an atypical picture of the disease. We encountered erroneous diagnoses in 81.3 per cent of patients with this anomaly.

Most frequently dystopic kidneys were mistaken for appendicitis. We encountered 92 such patients; they had all been subjected to unnecessary appendectomy and two, in addition, had undergone diagnostic laparotomy. After appendectomy the symptoms of the diseases remained or recurred after a lapse of time: it was only after this that the patients were sent for urological examination which helped to make the proper diagnosis of the dystopic kidney.

Urological examination was undertaken after appendectomy because urine tests showed abnormal values, a fact noted but neglected prior to operation. The following case is given in illustration.

G., a 32-year-old woman, was admitted to the clinic on March 13, 1972 complaining of pain in the right abdomen. Three weeks earlier she had undergone appendectomy. The abdominal pain was not relieved by the operation, however, and the patient's condition did not improve. Urinalysis: erythrocytes up to 30-40 in field of vision, protein up to 0.066 pro mille. Urine tests before operation showed similar values. Due to the results of these postoperative tests a kidney disease was suspected and the patient was sent for urological examination. Abdominal palpation illicited tenderness medial to the fresh postoperative scar. Pasternatskii's sign was positive on the right. Excretory urography showed the left renal pelvis and ureter on the radiographs; the right pelvis could not be seen but the right ureter was filled with the contrast medium. Retrograde pyelography on the right demonstrated an anomalous renal pelvis above the right ileosacral articulation, with its calyces facing inward and a short ureter. No signs of disease were found. A diagnosis of iliac dystopia of the right kidney was made.

The need for an urological examination usually becomes evident during an operation, when a tumour resembling a kidney is found in the retroperitoneal space.

S., an 18-year-old woman, was admitted to the clinic on April 18, 1962. She complained of dull pain in the right iliac region, where a tumour was palpated. An appendectomy had previously been performed. No changes were found in the removed vermiform appendage, but a tumour resembling a kidney was found in the retroperitoneal space to the right of the spine. The patient was sent for urological examination. On inspection a fresh postoperative scar was seen in the right iliac region. Below the scar and slightly toward the midline a smooth and immobile tumour was palpated. It was of hard elastic consistency and measured 7×9 cm. Retrograde pyelography and diagnostic pneumoretroperitoneum disclosed crossed dystopia of the kidney without fusion.

The diagnostic errors in these cases were due to the atypical picture of the disease (the unusual position of the tumour and haematuria which were noted only after operation). As can be seen from some of our cases, however, the unjustified operation could have been avoided if the physician had noted and correctly responded to the abnormal values of the urine tests. The presence of pus or blood in the urine indicates the need for urological examination.

Kh., a 30-year-old woman, was admitted to the clinic on April 12, 1952. She complained of attacks of pain in the right iliac region, where a tumour was palpated. Two weeks earlier she had undergone appendectomy during which a tumour resembling the kidney was found in the retroperitoneal space. The patient was sent for urological examination. A paramedian scar was seen to the right of the umbilicus. A hard tender tumour 8×8 cm in size was palpated under it. Urine test: protein 1.65 pro mille, leucocytes and erythrocytes covering the entire field of vision. Retrograde pyelography showed the right renal pelvis with amputation of the middle and lower calyces typical of dystopia. A diagnosis of tumour of a dystopic kidney was made. Nephrectomy was performed. Pathohistological diagnosis: trabecular adenocarcinoma of a dystopic kidney.

Urine tests very often show abnormal values in cases of urolithiasis. Scout radiographs of the abdominal cavity can help in making a proper diagnosis in this case. Urinary tests and X-ray examination are not undertaken, however, or their results are disregarded, which leads to unwarranted operations.

K., a 27-year-old woman, was admitted to the clinic on June 12, 1959 from the surgical department after appendectomy. She complained of aching pain in the right abdomen. Urinalysis: protein up to 0.1 pro mille, leucocytes 15 to 20 and erythrocytes 3 to 5 in field of vision. A scout radiograph showed the shadow of a calculus 1×0.8 cm in size in the lower third of the right ureter. Excretory urography and retrograde pyelography demonstrated lumbar dystopia of the right kidney. The calculus was removed by ureterolithotomy.

Chromocystoscopy may be of great assistance in the differential diagnosis of appendicitis and urolithiasis. It should be used in doubtful cases of abnormalities in the urine or an unusual abdominal tumour.

T., a 24-year-old man, was admitted to the clinic on October 4, 1955, complaining of attacks of acute pain in the right abdomen. He had undergone appendectomy but three days later again experienced sharp abdominal pain. He was sent for urological examination. Palpation on the right of the umbilicus and in the region of the fresh postoperative scar was painful. Pasternatskii's sign was positive on the right. Urinalysis: protein up to 0.99 pro mille, leucocytes cover the entire field of vision, erythrocytes 15 to 20 in field of vision. The shadow of a calculus 1.5×1.0 cm in size was seen in the upper third of the ureter. Excretory urography demonstrated a picture characteristic of lumbar kidney dystopia. The calculus was removed by ureterolithotomy.

Sometimes an erroneous diagnosis of appendicitis is made when the patient has not been subjected to a thorough objective examination. Examination through the vagina or rectum enables one, in some cases, to palpate a tumour in the minor pelvis resembling a kidney. This should oblige the surgeon to conduct a urological examination.

Sh., a 27-year-old woman, was admitted to the clinic on January 4, 1964 complaining of pain in the right abdomen. Prior to this she had undergone appendectomy in the district hospital which, however, did not relieve the pain. Abdominal palpation illicited tenderness in the right iliac region. Vaginal examination helped to determine a smooth tumour in the minor pelvis. The findings of retrograde pyelography confirmed the diagnosis of a pelvic dystopic kidney.

Diseases of a dystopic kidney sometimes give a picture of ileus. We encountered this in pyelonephritis of a dystopic kidney. The patient was subjected to unjustified laparotomy during which the dystopic kidney was found. Diagnostic laparotomy in such cases makes the patient apprehensive of another operation which is in fact really necessary.

An erroneous diagnosis could have been avoided if the abnormality in the urine and the presence of an abdominal tumour of uncertain localization had been properly interpreted. This should focus the attention of surgeons on the study of the urinary organs, the anomalies of which may cause effaced forms of a disease resembling the clinical picture of ileus.

Diseases of dystopic kidneys are often mistaken for tumours in the abdominal organs. Even now unjustified diagnostic laparotomy is performed because of suspected abdominal tumours, although it would be more correct to determine precisely the position of the tumour in relation to the abdominal organs and carry out urological examination before the operation.

I., a 23-year-old woman, was admitted to the clinic on May 18, 1963, for urological examination after laparotomy. The operation had been performed in the surgical department for an abdominal tumour. During the operation the tumour was found to be in the retroperitoneal space. On examination in the clinic a rounded tumour was palpated in the right iliac region. Blood and urine tests showed normal values. The diagnosis of iliac dystopia of the right kidney was made by means of pyelography. No treatment was required.

The symptom common to all these patients was an abdominal tumour of unusual localization which showed no changes typical of new growth in X-ray examination of the stomach and intestine. The atypical symptoms of these relatively rare kidney anomalies and the biased attitude of surgeons seeking to find a surgical disease in their specialized area lead to diagnostic errors and unjustified surgical intervention. When a tumour of unrecognized origin is found in the abdomen, one must, before all else, presume disease of an anomalous kidney and perform urological examination without fail.

In one of our cases a pelvic dystopic kidney was mistaken for torsion of an ovarian cyst. Timely urological examination helped to avoid the error. The examination was made because a dystopic kidney was suspected since the tumour was behind the rectum and shaped like a flat cake.

Now and again errors of an opposite kind are made, when anomalies of the female genitals or diseases of the abdominal organs are taken for a renal anomaly. This mistake leads to a delay in the necessary operation, which aggravates the disease. In one of our cases a haematometra was mistaken for a dystopic kidney. The absence of a ureteral orifice on the left side and lack of functioning and shadow of the kidney here in diagnostic pneumoretroperitoneum provided grounds for presuming aplasia of the kidney. However, coincidence of the ailment with the menstrual period, the resilient elongated tumour palpated on the anterior surface of the rectum and another tumour felt above the pubis

should have suggested that this was a case of double uterus and blind-end vagina. The blood accumulated during menstruation filled and distended the accessory vagina and caused pain. The diagnosis of duplicate uterus and duplicate vagina with haematometra of one of them was in fact confirmed during operation. The accessory uterus and vagina were resected.

In summing up it should be noted that urological examination of these patients was usually undertaken when abnormalities were found in the urine or a tumour of unclear localization was palpated or found in the abdomen during operation. Unjustified operations were carried out because of deficient examination. Erroneous diagnosis and unjustified operations were made because diseases of anomalous kidneys are a rare occurrence and often follow an atypical course. The underestimation of the results of urine tests, scout radiographs of the abdominal cavity and chromocystoscopy was of no small significance. A tumour palpated in the abdomen or any abnormalities of the urinary system indicate the need for preoperative urological examination. Our experience confirms that timely urological examination helps to make the proper diagnosis.

It is necessary to draw the attention of surgeons to this problem. They should bear in mind that in cases of suspected appendicitis, ileus or tumour of the abdominal organs the problem may in fact be a disease of a dystopic kidney producing a similar clinical picture. In unclear cases it is necessary to conduct a urological examination before undertaking a diagnostic operation. In emergency surgery the results of urine tests, scout radiographs of the abdominal organs and chromocystoscopy should be carefully analysed. Proper interpretation of the findings of these examinations will exclude unfortunate errors and unjustified operations.

During urological examination dystopic kidneys have to be differentiated from fused kidneys, nephroptosis and certain renal diseases (tumours, hydronephrosis, urolithiasis, tuberculosis) in which deformities of the urinary tract are encountered.

Bilateral dystopia of the kidneys is easily distinguished from fused kidneys by means of the methods discussed above. The difficulties of diagnosis are solved by telepyeloscopy and pneumoretroperitoneum. Dystopic kidneys are easily drawn apart under the screen and the image of each kidney is seen separately.

In nephroptosis the mobile kidney has pelves which normally extend inward from the calyces. This is seen well with excretory urography. If the function of the kidney is diminished or the pelves cannot be seen against the background of the pelvic bones the upper urinary tract may be clearly demonstrated by means of retrograde pyelography. In the rare doubtful cases dystopia

of the kidney can be verified by means of renal angiography showing short vessels originating from the aorta below the normal level for kidneys.

The unusual position of the calyces of a dystopic kidney can sometimes be mistaken for a filling defect in them in tumour of the kidney or for deformity of the upper urinary tract in a case of tuberculosis or urolithiasis. Such an error occurs only when the physician is not acquainted with the X-ray pattern of the upper urinary tract of a dystopic kidney. Experience in the interpretation of these radiographs enables one to avoid such an error easily.

Peculiar features of the symptomatology and diagnosis of diseases of dystopic kidneys. Dystopic kidneys are predisposed to certain diseases. Our experience shows that hydronephrosis, nephrolithiasis and pyelonephritis are encountered most frequently. In a group of 162 patients with dystopic kidneys 22.3 per cent had hydronephrosis, 11.7 per cent nephrolithiasis, and 11 per cent pyelonephritis. These diseases together accounted for 45 per cent of cases. The other diseases are encountered less frequently. We observed hypernephroma of a dystopic kidney in 2.9 per cent and nephroptosis in 5.8 per cent of cases.

Gimpelson (1949) found pathological mobility of a dystopic kidney particularly often, in 16 patients. Other diseases were encountered in 18 patients (nephrolithiasis in 13, pyonephrosis in one, tuberculosis in one, and tumour of the kidney in one). Thompson and Pace (1937) reported that among 97 patients with a dystopic kidney 15 had hydronephrosis, 20 pyelonephritis, 7 nephrolithiasis, 2 tuberculosis of the kidney and one had tumour of the kidney.

Sokolov (1928) claims that dystopic kidneys are affected by diseases in 76 per cent of cases. In 7 of 42 cases diseases of a dystopic kidney were the cause of death. Aizenshtein (1937) found diseases of the anomalous kidney in 30 per cent of patients with renal dystopia and Antonov (1964) in 54 per cent.

The special predisposition of a dystopic kidney to retention diseases is linked first of all with its unusual position below or above the normal level. The unprotected position of the kidney, closely surrounded by other organs, is conducive to compression and injury. The origin of the ureter from the middle part of the kidney, and its high, sharply angled origin from the pelvis create favourable conditions for urinary stasis and disturbed urodynamics in the upper urinary tract. Moreover, the ureter is often compressed by the accessory vessels often encountered in a dystopic kidney.

Thus, a dystopic kidney is undoubtedly predisposed to the development of diseases such as hydronephrosis, nephrolithiasis

and pyelonephritis. Other diseases are evidently encountered in this renal anomaly no more frequently than in normally developed kidneys.

Hydronephrosis. Hydronephrotic transformation of a dystopic kidney occurs mainly when the ureter arises from the pelvis at a high level and at a sharp angle and when the prepelvic segment of the ureter is compressed above the lower pole of the kidney. Periureteritis and compression of the ureter by accessory vessels which very frequently occur in this renal anomaly are other contributing factors. The remaining causes, such as a fixed ureteral kink or ureteral strictures, are rare in our experience.

The symptomatology of this disease in a dystopic kidney has no special features. The clinical picture, however, does not conform to the usual clinical picture of hydronephrotic transformation of the kidney.

It is our experience that all patients with hydronephrotic transformation of a dystopic kidney can be separated into the following four groups according to the clinical picture: (1) asymptomatic; (2) with symptoms of kidney dystopia; (3) with symptoms of kidney dystopia and disease; (4) with symptoms of the disease. The pathological condition is rarely a chance finding at an examination. We had only one patient with an asymptomatic course of this disease in a dystopic kidney. He had been admitted to the clinic for a planned follow-up examination. Patients with manifestations only of the renal anomaly are encountered more often: there were five in our practice. The remaining patients applied for medical advice with symptoms of the disease or with symptoms of the disease and the renal anomaly.

The most frequent manifestation in our cases was pain felt in different parts of the abdomen and rarely in the small of the back. It is difficult to attribute this pain to a renal disease and it is often accepted as a sign of various diseases of the abdominal organs.

The pain is usually dull and continuous or occurs at intervals. It is rarely sharp and rarely comes as an attack. Pain in lumbar kidney dystopia has the character of renal colic with its typical manifestations.

The unusual localization of the pain and the frequently observed dyspeptic disorders result in hydronephrotic transformation of a dystopic kidney being often mistaken for a gastro-intestinal disease, for which long-term treatment is applied with no effect.

P., a 23-year-old woman, was admitted to the clinic on October 21, 1961. She complained of dull pain in the region of the umbilicus and lower abdomen, nausea, eructation, constipation and, at times, abdominal distension. She had been treated unsuccessfully for three years for gastritis and later for colitis. A year previously she had undergone

appendectomy. The condition did not improve and she lost weight. On examination an elastic tumour was found in the abdomen in the right iliac region. It was suspected that the patient had a dystopic kidney and she was sent for urological examination. Blood tests showed no abnormalities. Urinalysis: protein 0.66 pro mille, leucocytes 30 to 40 in field of vision, erythrocytes 3 to 4 in field of vision. X-ray examination enabled the physician to diagnose hydronephrotic transformation of the right iliac dystopic kidney. Nephrectomy was performed. The patient recovered.

In this case an abdominal tumour found near to the midline served as the reason for the urological examination. It helped us make the correct diagnosis and apply the necessary treatment.

The occasional disappearance of the tumour followed by polyuria in evacuation of the cavity of the kidney is characteristic of hydronephrotic transformation of the kidney. The appearance of the tumour in the abdomen is sometimes attended by elevation of body temperature, a chill, and pain. We encountered this in several patients. A dystopic kidney that undergoes hydronephrotic transformation is palpated particularly often. The tumour in such cases is large, painful, elastic and, most important, it lies near the midline of the body and is poorly mobile.

Dysuric disorders, which are usually caused by secondary infection or are of a reflex character, are the most frequently encountered signs prompting the physician to conduct a urological examination, as a result of which a dystopic kidney with hydronephrotic transformation is revealed. Dysuric disorders, haematuria, pyuria and pseudoproteinuria, in addition to abdominal pain and an abdominal tumour palpated near the midline, suggested hydronephrotic transformation of a dystopic kidney.

S., a 17-year-old boy, was admitted to the clinic on September 28, 1971. He complained of frequent and painful micturition and dull pain in the abdomen to the left of the umbilicus, where he himself could feel a tumour. Blood tests showed normal values. Urinalysis: protein 0.99 pro mille, leucocytes covering the entire field of vision, erythrocytes 10 to 15 in field of vision. Chromocystoscopy: hyperaemia in the area of the left ureteral orifice, indigo carmine was discharged from the right orifice in the third minute and from the left in the sixth minute. Excretory urography showed a distended shadow of the pelvis immediately above the ala of the ilium with calyces facing the vertebrae. The findings of retrograde pyelography confirmed the diagnosis of hydronephrotic transformation of the left lumbar dystopic kidney.

There are reports in the literature on rupture of a hydronephrotic dystopic kidney into the abdominal cavity (Dobrovolskaya, 1925; Kubarikov, 1961). In such cases the symptoms of an acute surgical disease of the abdominal organs come to the forefront. The diagnosis of hydronephrotic transformation of the dystopic kidney is made during the operation, since the symptoms of the disease are vague.

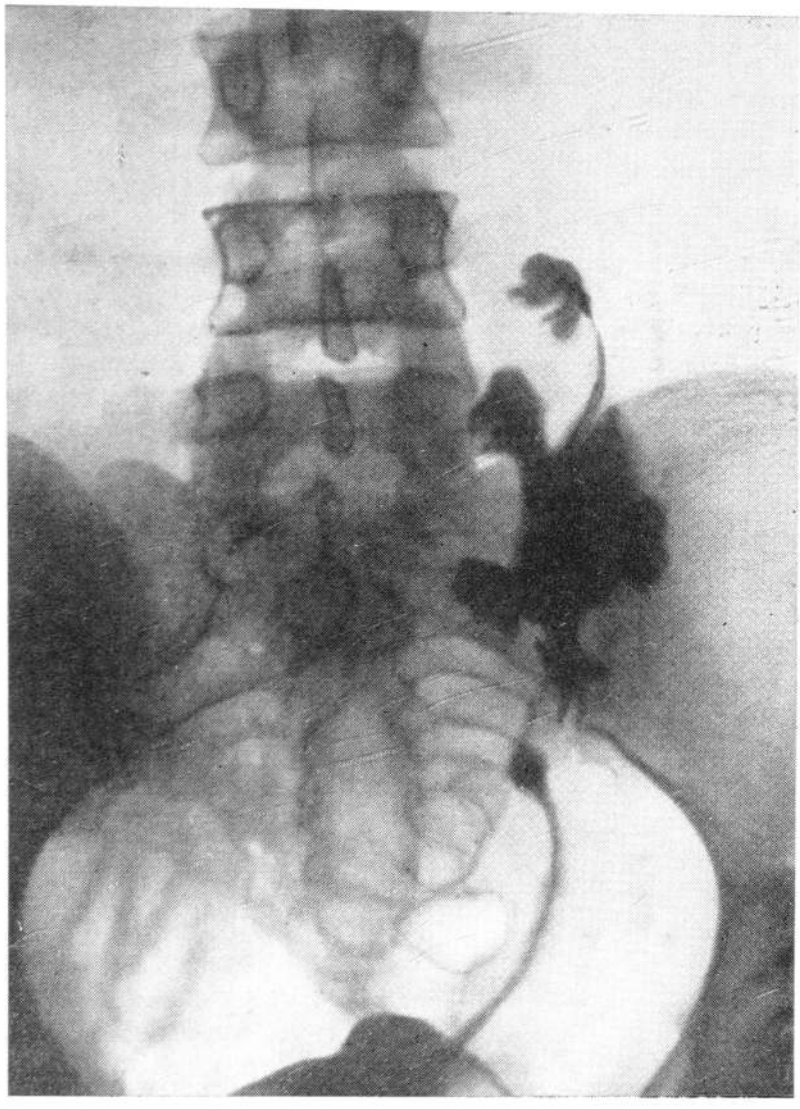


Fig. 85. Case R. Hydronephrotic transformation of an iliac dystopic kidney. Retrograde pyelography

Hydronephrotic transformation of a dystopic kidney follows a chronic clinical course. Some patients date its symptoms back to childhood and are unsuccessfully treated for different diseases over many years. In some cases the disease is manifested at a later age. The aggravation of the disease from time to time makes these patients seek medical advice more and more often. It is during one of these examinations that the disease and renal anomaly are recognized.

The diagnosis of hydronephrotic transformation of a dystopic kidney is based on the symptoms discussed above which lead

one to suspect a kidney disease or anomaly. The diagnosis may be verified or ruled out by X-ray and other methods of examination. The value of the various methods is discussed earlier in this chapter.

Retrograde pyelography yields convincing data for diagnosing the anomaly and the disease. The pyelograms (Fig. 85) show a distended pelvis whose calyces are directed at the vertebrae. The pelvis has a low position and the ureter is short. But it is difficult to suspect kidney dystopia in cases of severe hydronephrosis, when the calyces merge with the pelvis. In these cases only the low position of the renal cavity and the short ureter indicate that this is a case of dystopic kidney. Retrograde pyelography helps to ascertain the exact causes of the hydronephrotic transformation of the kidney.

In hydronephrotic transformation the image of the dystopic kidney on the radiographs is larger than normal. This can be taken as indirect proof of the disease, and can be seen with the aid of tomography or diagnostic pneumoretroperitoneum. The combination of these methods with excretory urography or retrograde pyelography helps to assess the amount of preserved renal parenchyma.

When it is difficult to make a diagnosis, the renal anomaly and the disease can be recognized by means of renal angiography. A pattern poor in vessels which are thin and tortuous indicates hydronephrotic transformation of the kidney while the low origin of the vessels from the iliac artery is characteristic of renal dystopia. Renal angiography yields valuable data and merits application before surgery on such a kidney. The results of this examination help in the choice of operation.

Cineurography and urokymography provide information on the function of the upper urinary tract in such cases. Weakening or absence of pelvic contractions is usually noted in hydronephrotic transformation. The amplitude of the waves depends on the degree to which the muscular wall is preserved. In many cases some parts of the ureter cannot be seen during contraction of its prepelvic segment, and the ureter is displaced laterally, which indicates that it is compressed above the pole of the dystopic kidney.

Nephrolithiasis. The clinical picture of renal and ureteral stones in cases with dystopic kidney is marked by certain peculiarities. In some cases only symptoms of the disease form the clinical picture; in others both the symptoms of the disease and the symptoms of the anomaly are manifest, the former predominant over the latter.

All patients with nephrolithiasis of a dystopic kidney complain of pain. It often occurs in attacks which are followed by pe-

riods of dull pain. The localization of the pain is unusual for one caused by a kidney. It is felt in different parts of the abdomen and rarely (in lumbar kidney dystopia) in the small of the back. This localization suggests a disease of the abdominal organs, which is why many patients are often unsuccessfully treated for years on end for gastro-intestinal diseases or diseases of the biliary tract. In one of Topchibashev's (1957) cases, calculi in a dystopic kidney simulated the clinical picture of ileus.

A smooth elastic and immobile tumour palpated near the vertebrae suggests a disease of a dystopic kidney. Urological examination helps to verify the diagnosis.

On scout radiographs calculi in a dystopic kidney can be mistaken for calculi in the ureter due to their low position. Large calculi seen in a low position on the radiographs should suggest the possibility of a renal anomaly, particularly a dystopic kidney with numerous or coral calculi filling its pyelo-calyceal system.

The image of the urinary tract produced in excretory urography and retrograde pyelography helps in the correct diagnosis of the renal anomaly and in an assessment of the character of the functional and morphological changes in the kidney. Renal angiography yields valuable information in cases in which diagnosis is difficult.

Pyelonephritis. The clinical picture of pyelonephritis of a dystopic kidney is made up of symptoms of the disease and of the renal anomaly or of symptoms of the disease alone. Pain is the main sign. It is atypical and localized in the abdomen, rarely in the small of the back. The palpation of a tumour in the abdomen with its characteristic signs suggests a dystopic kidney. The other symptoms of the anomaly (dyspeptic disorders) create difficulties in the diagnosis. Such patients receive treatment for erroneously diagnosed diseases of the abdominal organs with no effect and undergo unjustified operations.

The characteristic abdominal tumour, the occasionally encountered dysuric disorders or pain in the small of the back and pyuria are the symptoms which prompt the physician to perform a urological examination. It reveals the dystopic kidney and the disease. In case of fever, dysuric disorders and a chill, i.e. when there is a clinical picture of an inflammatory disease of the urinary system, it is assumed that the patient has pyelonephritis and the anomalous kidney becomes a chance finding during urological examination. Excretory urography, renography, scanning of the kidneys, and renal angiography help to make the proper diagnosis in difficult cases.

Complications in pyelonephritis of a dystopic kidney have been described in the literature. They present even greater difficulties in making a diagnosis and are often mistaken for dis-

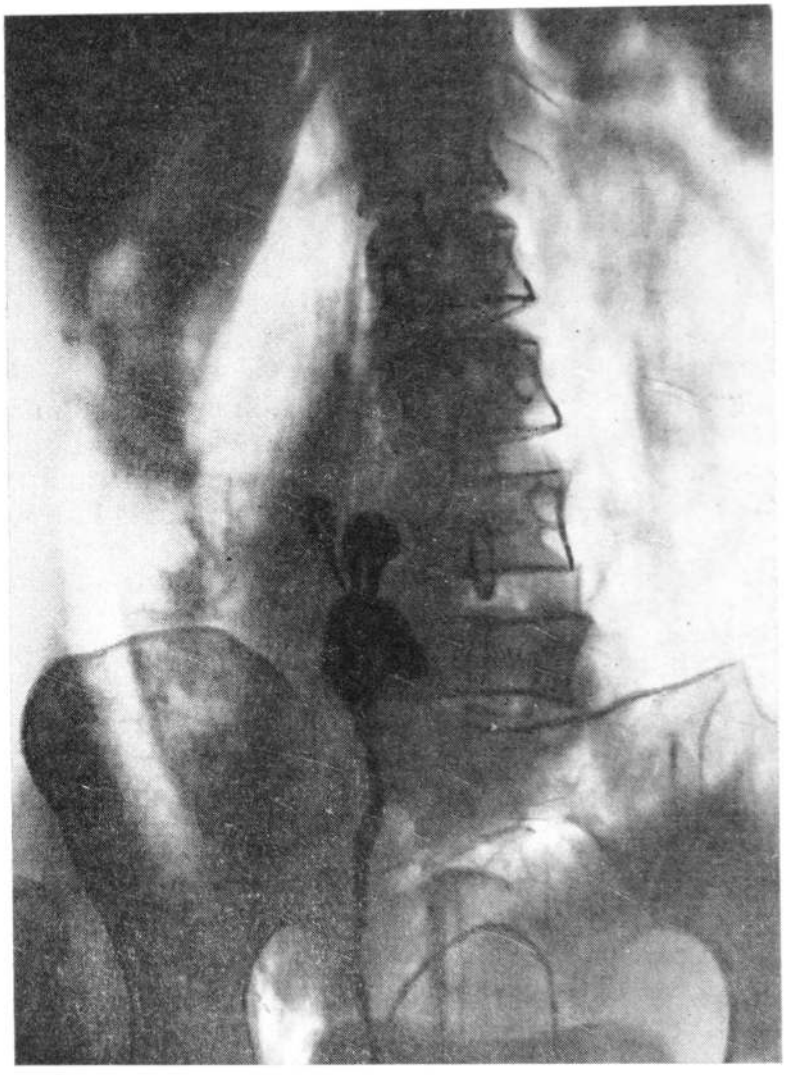


Fig. 86. Case Kh. Hypernephroma of an iliac dystopic kidney. Retrograde pyelography

eases of the abdominal organs. We had a patient with acute purulent pyelonephritis of an iliac dystopic kidney who had been treated for an abscess of the intestinal loops.

Tumour of dystopic kidney. There are occasional reports in the literature of adenocarcinoma, hypernephroma and cancer of the pelvis of a dystopic kidney (Borchardt, 1932; Lee, 1949; Caine, 1956; Raevsky, 1957; Castro and Ashken, 1972). We had five patients with hypernephroma of an iliac dystopic kidney. Pain, a palpable tumour, and profuse intermittent haematuria are the main symptoms of a tumour of a dystopic kidney. It would seem that this triad of symptoms should help in the time-

ly diagnosis of a renal tumour. Here too considerable diagnostic difficulties arise. Patients are sent to a urological clinic late, often after they had been subjected to laparotomy in the surgical department.

Kh., a 30-year-old woman, was admitted to our clinic after an appendectomy. She had complained of attacks of pain in the right iliac region. The tumour palpated here was taken for appendicular infiltrate. Urological examination made after the operation helped in the diagnosis of tumour of a dystopic kidney. Retrograde pyelography (Fig. 86) showed a distorted pelvis on a level with the sacroiliac articulation and the fifth lumbar vertebra, two calyces directed upwards, and a short ureter.

The unusual localization of the pain leads to diagnostic errors. It is felt to the right or left of the umbilicus and in the iliac region. The abdominal localization of the tumour so uncommon for a kidney is also the cause of it being mistaken for an abdominal new growth or, as in the case described above, for an appendicular infiltration. Haematuria alone in this triad of symptoms is characteristic of a renal disease. The other symptoms of the kidney anomaly just confuse the clinical picture.

A diagnosis of a renal anomaly and tumour is based on the findings of retrograde pyelography. The low position of the renal pelvis, calyces directed at the vertebrae, the short ureter and, in addition, a filling defect in the calyces or pelvis enable the physician to make a correct diagnosis of both the anomaly and the disease. Sometimes the interpretation of such radiographs is difficult. The filling defect in the calyces may be attributed to a peculiar structure of the urinary tract of an anomalous kidney. In this case, however, the clinical picture of the renal tumour helps one avoid mistakes.

Renal angiography should be used in cases which are difficult for diagnosis. On the radiographs a renal area with a thick vascular network corresponds to the tumour.

Tuberculosis. Only rare cases of tuberculosis of dystopic kidneys are cited in the literature. The clinical picture is usually made up of symptoms of the disease and symptoms of the renal dystopia. As in the diseases of dystopic kidneys described above, abdominal pain and a tumour in the abdomen which is an uncommon site for a kidney create difficulties of diagnosis. Such patients are sent for examination because of a suspected abdominal tumour.

A diagnosis of tuberculosis of a dystopic kidney is based on the results of retrograde pyelography, which demonstrates destructive changes in the upper urinary tract and characteristic signs of kidney anomaly.

Among other methods of examination it is necessary to emphasize the significance of medical history, tests for pus and

Mycobacterium tuberculosis in the urine, the presence of specific tubercles on the mucous membrane of the urinary bladder, and also functional and morphological changes in the kidney revealed by chromocystoscopy, excretory urography, renography, scanning of the kidneys and renal angiography.

Nephroptosis. Gimpelson (1949) claims that pathological mobility of the dystopic kidney is often encountered and produces a clinical picture of nephroptosis. Even mild mobility of an anomalous kidney held by several vessels on the nerve plexuses causes pain when the person walks or performs physical work.

Our experience, however, does not confirm the high incidence of this disease in dystopic kidneys. Patients who complained of pain associated with dyskinetic disorders and the first manifestations of urinary stasis in the anomalous kidney were evidently included in this group.

The clinical picture of this disease is also made up of the symptoms of the disease and the symptoms of the renal anomaly. But the manifestations of the disease are more marked. Excretory urography, renal angiography and cineurography are important in making the diagnosis.

Trauma. There are occasional reports in the literature on damage to a dystopic kidney (Zalessky, 1862; Kuzmenko, 1936; Gorshkov, 1966; Konstantinovsky and Kuznetsov, 1967); it is highly probable that it is damaged more frequently because of its unprotected low position.

Patients with trauma of a dystopic kidney are usually admitted to the clinic with an erroneous diagnosis and the damage to the kidney is revealed during diagnostic laparotomy. The clinical picture simulates that of injury to the abdominal organs. It is very difficult to arrive at the correct diagnosis, which is only possible when the surgeon bears in mind that a trauma could have been inflicted on a dystopic kidney and, on finding a low abdominal tumour, undertakes a urological examination. Among the methods of examination chromocystoscopy, renography and excretory urography merit attention in such acute cases.

Treatment of diseases of a dystopic kidney. People with dystopic kidneys that have no clinical manifestations or diseases require no treatment. Indications for the removal of an anomalous organ occur only in rare cases when a dystopic kidney in the pelvis hinders delivery.

Non-operative treatment in the initial stage of disease and disorders does not usually differ from that applied to different diseases of a normally developed kidney. Because of urinary stasis in the pelves catheterization of the pelvis has to be performed for its adequate evacuation. Thompson and Pace (1937) particularly recommended this type of treatment. It is our expe-

rience, however, that in most cases non-operative treatment brings only temporary relief. Non-operative treatment is recommended if the dystopic kidney causes only mild pain which is relieved for considerable lengths of time by therapeutic measures. In frequent exacerbations of the disease resulting in loss of working capacity, such treatment is insufficient. It should also be emphasized that non-operative treatment does not prevent the progress of a disease of a dystopic kidney.

Z., a 24-year-old man, was admitted to the clinic on January 27, 1973, complaining of sharp pain in the right abdomen, nausea, constipation and frequent micturition. He had been ill for five years. At first the attacks of sharp pain, dyspeptic disorders and dysuria had troubled the patient no more than once a year. In the year before admission to the clinic the attacks of pain occurred every month. Working capacity was lost. Palpation revealed an elastic, painless, and poorly mobile tumour in the right iliac region. Blood tests showed no abnormalities. Urinalysis: protein up to 0.66 pro mille, leucocytes and erythrocytes up to 10-15 in field of vision. Ambourget's test confirmed that the patient had pyuria. Active leucocytes were revealed. X-ray examination showed the pelvis of a dystopic kidney on the right above the sacroiliac articulation; its calyces were pointed at the vertebrae and the ureter was short. The pelvis and the calyces were distended. Comparison with radiographs of this patient taken in previous years showed that the cavities of the pelvis and calyces of the dystopic kidney were gradually distended with time. A diagnosis of hydronephrotic transformation and pyelonephritis of an iliac dystopic right kidney was made. Surgery was recommended.

It can be seen from this case that progression of the clinical manifestations of the disease ruled out non-operative treatment.

Surgery is the principal method for the management of diseases of a dystopic kidney.

Indications for surgery. In cases of dystopic kidneys uncomplicated by disease surgery is indicated when, because of continuous dull pain or frequent attacks of sharp pain, the patient is incapable of work. One must agree with Punin (1946) that an operation is indicated if pelvic dystopia of the kidney prevents delivery.

The patient's age and general condition, the condition of the cardiovascular system, the degree and character of the clinical manifestations of the disease, the functional and morphological changes in the kidney, the presence and virulence of infection, and the causes of the disease all have a direct bearing on the indications for surgery. Urinary stasis in the pelvis of a dystopic kidney complicating the course of the disease has to be taken into account in considering the indications for operation, which is why we distinguish indications for surgery in diseases of a dystopic kidney with urinary stasis in the pelves and indications for operations in other diseases.

On the basis of our personal experience, we believe that sur-

gery should be performed on all patients with hydronephrotic transformation of the kidney, when the disease produces marked clinical symptoms or is progressive, which can be revealed during regular follow-up, and in cases of concomitant infection and disorders of renal function. Judging from the data of Ostropolskaya (1970), temporizing tactics in hydronephrotic thansformation of the kidney in children often leads to infection of the kidney and its rupture through the injuries often suffered by children. For this reason we consider such tactics unwarranted.

We consider an operation indicated in nephrolithiasis when the patient is incapacitated by frequent renal colic, and also when there are calculi in the kidneys and ureters with inflammatory complications and abnormal renal function. The obstacles to the flow of urine in the prepelvic segment of the ureter of a dystopic kidney hamper the elimination of calculi in many cases. This is why, when there are morphological and functional changes in the kidney, or when concomitant pyelonephritis develops, it would be wrong to wait too long for the spontaneous elimination of the concrement.

Operative treatment is indicated in cases of recurrent pyelonephritis of a dystopic kidney when there is stasis of urine in the pelvis, which is a frequent occurrence. Surgery is necessary in complications of pyelonephritis (pyonephrosis, acute purulent pyelonephritis, etc.).

In all other diseases not associated with urinary stasis in the pelves, the indications for surgery do not differ from those in diseases of normally developed kidneys.

Isakova et al. (1967) believe that absolute indications for operations on children are severe complications which drastically impair the flow of urine and which are marked by far advanced hydronephrotic transformation. Surgery is likewise necessary in cases of an intrarenal type of pelvis with hydronephrotic transformation, because in such cases the function of the organ is quickly lost. Operation is indicated in the initial stages of hypertensive disease. The authors consider that in moderate complications of disturbed urinary flow and mild secondary anatomical and functional changes the indications for operation are of a relative character.

Contraindications for operative management of diseases of dystopic kidneys lack any special features. In such cases, just as in cases of fused kidneys, immobility of the kidney cannot be regarded a sign of an inoperable tumour, because a dystopic kidney is usually firmly attached to its seat.

Choice of operative method. As one can see from the above, a dystopic kidney with no marked signs of disease can still cause pain and disorders. Depending on the interpretation of the causes

of these disorders there are several methods of treatment.

There is an opinion that pain in people with dystopic kidneys is caused by compression of the nerve ganglia and plexuses by the abnormally attached organ, to which sclerotic paranephritis may be conducive. This is why many surgeons perform nephrectomy if pain incapacitates the patient (Lukina, 1911; Sauer, 1926; Punin, 1946; Kapustin, 1962). This operation, naturally, is possible only if the contralateral kidney is healthy. Gimpelson (1949) thinks that this operation is necessary in pelvic kidney dystopia even if the function of the organ is good and there is no infection. Conservative surgery is ineffective in this case. Moreover, the anomalous kidney may prevent delivery of a baby.

Gotlib (1925) and Gimpelson (1949) attributed pain in cases of dystopic kidney to the abnormal mobility of the organ. They claim that if pain is associated with abnormal mobility of the anomalous kidney, operation for nephropexy is indicated. It corrects the excessive mobility of the kidney, changes its position and thus removes the pressure of the kidney on the adjacent nerve plexuses and organs, and improves the flow of urine from the pelvis. But if nephropexy cannot be performed because of a short renal pedicle, they recommend nephrectomy.

A variant of this operation is that performed by Braitsev (1926) for a dystopic kidney. After exposure the kidney was turned by 180 degrees so that the ureter was medial to it, after which nephropexy was carried out. Braitsev claims that this operation eliminates stretching of the renal vessels, the main cause of pain. The operation was used in one of our cases.

For complete relief of pain in dystopic kidney Chaika (1929) performed denervation (he decapsulated the kidney and divided the nerves stretching along the artery and veins). This operation, however, had a short-lived effect and was not performed again.

Judging from the data in the literature (Brain, 1942; Gimpelson, 1949; Spigel, 1951) nephrectomy is used widely in the management of hydronephrotic transformation of a dystopic kidney. The operation is more readily undertaken on a dystopic kidney than in cases of hydronephrotic transformation of a normally developed kidney. The reason for this are the frequent recurrences of the disease after conservative surgery.

It is our experience that nephrectomy in hydronephrotic transformation of a dystopic kidney should be undertaken only in pelvic dystopia as well as in cases of marked morphological changes in the dystopic kidney and absence of function. Cases of solitary dystopic kidney and bilateral diseases with the function of the contralateral kidney diminished should be managed by an organ-preserving operation.

It is necessary to remove the obstacle to the flow of urine from the pelvis and restore the dynamics of its drainage by means of a plastic operation. The anatomical relationship of the kidney and its urinary tract should be restored as closely as possible to normal. Our experience shows that the frequent localization of the pelvis on the anterior surface of the dystopic kidney, the high and acutely angled origin of the ureter from the pelvis, and compression of the prepelvic segment of the ureter above the kidney are the principal causes of hydronephrotic transformation of a dystopic kidney. This is why a plastic operation cannot be restricted to correcting the stenosis in the prepelvic segment of the ureter and its compression by an accessory vessel. It is also essential to transpose the prepelvic ureteral segment below the pole of the kidney. This will correct the high origin of the ureter from the pelvis and compression of its prepelvic segment above the kidney. For this purpose we suggest resection of the lower pole of the dystopic kidney (Shimkus, 1969).

Sh., a 19-year-old woman, was admitted to the clinic on December 23, 1970 complaining of dull pain in the left iliac region. She had been ill for three years. The diagnosis of hydronephrosis of a left iliac dystopic kidney had been made earlier. Non-operative treatment proved ineffective. A smooth elastic and painless tumour was palpated in the left iliac region. Blood test showed normal values. Urinalysis: protein 0.66 pro mille, leucocytes 30-40 and erythrocytes 5-6 in field of vision. X-ray examination revealed a dystopic kidney with a characteristic urinary tract in the left iliac region. Its pelvis was distended. Pyelotelescopy and urokymography showed disturbed dynamics of the prepelvic segment of the ureter of the dystopic kidney and high origin of the ureter from the pelvis. The pelvis of the normally developed right kidney was also slightly distended. The function of both kidneys was quite satisfactory. The following diagnosis was made: hydronephrotic transformation of left iliac dystopic kidney, initial phase of hydronephrotic transformation of right kidney, pyelonephritis. Operation was performed on the left kidney, during which it was found that the hydronephrotic transformation had been caused by the high origin of the prepelvic ureteral segment from the pelvis, periureteritis, and the compression of the prepelvic segment of the ureter above the lower pole of the anomalous kidney. Resection of the lower pole of the dystopic kidney and plastic surgery of the pelvis after Foley were performed. The patient recovered.

Resection of the lower pole should be an indispensable component of a plastic operation on a dystopic kidney in cases of disturbed urodynamics in the prepelvic ureteral segment and urinary stasis in the pelvis, when one of the causes of these changes are compression of the ureter above the pole of the kidney and its high origin from the pelvis. Such reconstructive corrective operation improves the outcome of the treatment of diseases of dystopic kidneys.

Plastic operations without resection of the lower pole of the kidney are often unsuccessful. They may be recommended in

those rare cases when the ureter of a dystopic kidney lies to the side of the pole and does not arise from the pelvis at a high level.

Extra care should be taken in the management of hydronephrotic transformation of the kidney in children. One must agree with Ostropolskaya (1970) that nephrectomy in children is indicated in cases of total destruction of the kidney with loss of its function and in crossed and pelvic dystopia of the kidney with hydronephrotic transformation. In all other cases corrective operations are essential.

Nephrectomy is also widely used in cases of stones in a dystopic kidney. Gimpelson (1949) recommended this operation in cases of infected stones with diminished function of the kidney and distension of its cavities. If the renal parenchyma is preserved and there is no deep infection he believed pyelolithotomy could be applied.

The unfavourable anatomical features of a dystopic kidney often cause recurrences after organ-preserving operations on it. Here, just as in hydronephrotic kidney transformation, reconstructive operations with resection of the pole of the kidney are necessary in cases of high origin of the ureter from the pelvis and compression of its prepelvic segment above the kidney. This may prevent recurrences of diseases.

Excessive mobility of the dystopic kidney is an indication for nephropexy. This operation removes the symptoms of nephroptosis and is used quite widely (Gotlib, 1925; Punin, 1946; Gimpelson, 1949; Rabinovich, 1968). It is our experience, however, that undue preference for this operation is not warranted. Quite often, sooner or later, all the symptoms of the disease recur, pyelonephritis develops, and patients have to be operated on again for removal of the dystopic kidney.

K., a 43-year-old woman, was admitted to the clinic on March 23, 1964. She complained of continuous dull pain in the left abdomen and attacks of renal colic occurring now and then on the left side. Three years earlier she had undergone an operation for nephropexy on the left dystopic kidney. She felt well for a year after the operation, but then pain in the abdomen recurred and attacks of renal colic developed again from time to time. All the symptoms of the disease were more marked than prior to the operation. Urological examination showed that the kidney was fastened firmly in its seat and its mobility was restricted. Persistent pyuria and diminished function of the dystopic kidney were evidence of developing pyelonephritis. The pelvis of the kidney that had been operated on was moderately distended. There were no abnormalities in the other kidney. The clinical symptoms mentioned and incapacitation of the patient indicated removal of the dystopic kidney. The patient recovered. Pathohistological examination of the removed kidney showed pyelonephritis.

The methods of operative treatment for other diseases of dystopic kidney did not differ from those used in the same diseases of normally developed kidneys.

Special features of operative technique. The approach in operation on a dystopic kidney has some specific features. The deep localization of the anomalous kidney, which is covered by the peritoneum, its close contact with the large vessels, the great number of vessels, and the abnormal anatomy and poor mobility of the organ all create greater difficulties during operation than when dealing with a normally developed kidney. They can be overcome by an oblique extraperitoneal approach in operations on iliac or lumbar dystopic kidney. It is less difficult to operate on a pelvic or thoracic kidney through a transperitoneal or intrathoracic approach. Puterman (1949) notes that in repeated pyelolithotomy it is easier to remove the calculus transperitoneally. Because of the hazards of infecting the abdominal cavity and possible complications, this method should not be recommended for wide use.

In nephrectomy, the kidney should be exposed with great care and all the numerous vessels ligated, some of which, particularly those on the posterior surface, enter the parenchymal substance directly. Repeated nephrectomy is particularly difficult. During this operation the kidney has to be separated in the cicatricial tissue from the aorta, vena cava and iliac vessels, and numerous renal vessels, from which only the kidney's lateral border may be free, have to be ligated.

Organ-preserving operations on a dystopic kidney hardly differ from the commonly accepted procedures in the technique. They are performed without drawing out the kidney into the wound because it is firmly held in its seat by numerous vessels. The unusual position of the renal pelvis on the anterior surface of the organ makes anterior pyelolithotomy in nephrolithiasis easy. In hydronephrotic transformation of the kidney in children, Ostropolskaya (1970) recommends resecting the altered areas of the ureter and pelvis and preserving vascularization of the organ to the maximum. After the operation the kidney should be fastened in the most favourable position so as to avoid pathological mobility.

The great number of anatomical variants of dystopic kidneys rule out a standard approach to nephropexy. Gimpelson (1949) claims that in lumbar dystopia it is convenient to attach the kidney by its capsule to the twelfth rib, while an iliac kidney may be attached by the capsule to the iliac muscles. During this operation the dystopic kidney should be raised slightly and pulled laterally. Stretching of the vessels and the ureter during the procedure should be avoided. Stretching of the vessels causes circulatory disorders in the kidney and consequent pyelonephritis and associated symptoms of the disease which often make it obligatory to repeat nephrectomy.

Resection of the lower pole of the dystopic kidney in a corrective operation is performed by routine methods. It is important to resect the kidney to the lower border of the pelvis so as to transpose the prepelvic ureteral segment downward. If the length of the vessels and the shape of the kidney permit, the kidney may be turned with the pelvis facing medially and then fastened in this position. But this is not absolutely necessary. The great number of vessels and the flat shape of the dystopic kidney make this manipulation impossible in many cases. After resection it is sufficient to move the lower pole slightly laterally and fasten it to the muscle in this position.

Results of operative treatment. The immediate and early results of operations on dystopic kidneys are usually reported in the literature. The late results have been described only in individual cases, which does not give a good idea of the actual outcomes of these operations in general. This is why we studied the late results in our own cases. We assessed them as follows: excellent results (absence of complaints, restored working capacity, normal urine values, total restoration of renal function, restoration of normal volume of the renal pelvis and calyces); good (no complaints, restoration of working capacity, mild pyuria, improved renal function, reduced volume of the pelvis and the calyces); fair (subjective improvement, pyuria, no improvement in renal function, arrest of further destruction of the kidney); poor (persistence of complaints and pyuria, progressive deterioration of renal function, increased volume of pelvis and calyces, episodes of pyelonephritis, recurrences of stone formation).

As is noted in the literature, operation for nephropexy usually has good immediate and early results (Gotlib, 1928; Levitanus, 1935; Zakharov, 1967). Pain is relieved and the patient is able to resume work. Gimpelson (1949) obtained good results from nephropexy in most but not all of his patients. Punin (1946) claims that the best results were achieved in operations for lumbar and iliac renal dystopia, while in pelvic dystopia they were poorer, because sometimes the kidney cannot be drawn out of the true pelvis and attached at a higher level.

According to our experience, the late results of nephropexy on a dystopic kidney uncomplicated by disease are very disappointing. They were poor in three cases, fair in two and good only in one case.

Z., a 27-year-old woman, was admitted to the clinic on March 22, 1971. She complained of pain experienced now and then in the left half of the abdomen and dyspeptic symptoms. In the past year the pain had often troubled her and she could not work properly. Lumbar dystopia of the left kidney was diagnosed after urological examination. The kidney was mildly mobile. There were no symptoms of disease. Nephropexy was performed on the left dystopic kidney. The patient recovered. In the following six months she

felt well but then the pain recurred and became so intense that she could no longer work. The woman was again operated on. Nephrectomy was carried out. In the two years following the operation she considered herself healthy.

In this case the operation for nephropexy was unsuccessful. The kidney was slightly raised during it and fastened in that position. The vessels and ureter were not stretched.

In another of our cases nephropexy was performed after the dystopic kidney had been turned 180 degrees.

Ch., a 27-year-old woman, was admitted to the clinic on March 3, 1966. She complained of attacks of pain in the right abdomen and right lumbar region and dyspeptic and dysuric symptoms. Urological examination revealed bilateral lumbar kidney dystopia. Operation was performed during which it was found that the long vascular bundle of the right kidney stretched along its anterior surface. The kidney was turned 180 degrees, as a result of which the vessels were relaxed, and was also slightly raised and fastened in this position. The renal pelvis proved to be at the medial edge of the organ. The patient considered herself healthy for five years after the operation. Later dull lumbar pain developed. Regular check-ups revealed gradually progressing distension of the renal pelvis. In 1973 another operation for ureterolysis and nephropexy had to be undertaken because of hydronephrosis of the dystopic kidney. It was found that hydronephrotic transformation of the kidney was caused by stenosis of the prepelvic ureteral segment due to sclerotic periureteritis. The patient recovered.

In this case the poor results of the operation became evident five years after surgery. Stenosis of the prepelvic segment of the ureter developed because of sclerotic periureteritis. This complication may occur after any nephropexy.

Poor results from nephropexy are associated with progressive pyelonephritis, dyskinesia, and retention of urine in the renal pelvis because of the persisting compression of the prepelvic ureteral segment above the kidney or high origin of the ureter and stenosis of the ureter due to periureteritis. Since nephropexy often produces poor results and repeated nephrectomy has then to be performed later, the former should not be undertaken to relieve pain caused by the dystopic kidney. Nephrectomy is more reliable in such cases. We performed it to bring relief from pain caused by a dystopic kidney in five patients. All of them recovered and resumed work. Two of the five, however, developed a disease of the remaining kidney (pyelonephritis in one, the onset of hydronephrosis in the other) in the late postoperative period, which is why scrupulous follow-up examinations of such patients are essential so as to reveal and treat diseases of the remaining kidney in time. It is likewise necessary to search for other methods of operative treatment of disorders in cases of dystopic kidney.

The results of palliative operations in diseases of dystopic kidneys are often poor. In our experience disease recurred in

eight of 18 patients who underwent pyelolithotomy for nephrolithiasis of a dystopic kidney. In three of five plastic operations on dystopic kidneys, the late postoperative results were poor. In two of three of our cases nephropexy performed for nephroptosis of a dystopic kidney had poor results in the late period. The poor outcome of such operations is primarily due to the fact that the obstacle to the flow of urine from the pelvis remains because of the unfavourable anatomical features of the anomalous kidney.

Nephrectomy is the most commonly used operation in diseases of dystopic kidneys. Good immediate and occasionally late results were obtained with the operation by Osipov (1935) and Gimpelson (1949) in hydronephrotic transformation of the kidney, Thompson and Pace (1937) in complications of pyelonephritis, and by Nachman (1944) in tuberculosis. Balas et al. (1971) relieved a patient from high arterial hypertension in pyelonephritis by means of nephrectomy. Ostropolskaya (1970) observed good results from nephrectomy performed on children for hydronephrotic transformation of the kidney.

We studied the late postnephrectomy results in 23 cases of diseases of dystopic kidneys. They proved to be good and fair in 16 and poor in seven. The poor results were linked with diseases of the remaining kidney (hydronephrotic transformation, pyelonephritis and its complications, nephrolithiasis) and with metastatic spread of hypernephroma. The relatively large number of poor outcomes of nephrectomy calls for a cautious approach to the operation and restriction of the indications for it.

We had good results in three cases after corrective operations with resection of the lower pole of the dystopic kidney.

Z., a 21-year-old woman, was admitted to the clinic on October 11, 1969. She complained of continuous dull pain in the right iliac region which intensified now and again and was referred to the small of the back. The diagnosis of hydronephrotic transformation of the right iliac dystopic kidney was made from urological examination. The function of the kidney was satisfactory. It was found during the operation that hydronephrotic transformation of the dystopic kidney had been caused by the high origin of the ureter from the pelvis with compression of the prepelvic segment above the lower pole of the kidney and periureteritis. An operation for resection of the lower pole of the dystopic kidney and resection of the renal pelvis was performed. As the result of the manipulation the prepelvic segment of the ureter was displaced downward to a level below the pole of the kidney. The lower pole of the kidney was slightly drawn laterally and fastened in this position to the muscles. The patient recovered. She was followed up for a period of five years. The result was excellent.

We attribute the good results of these operations to the creation of an effective flow of urine from the pelvis of the dystopic kidney. These corrective operative interventions with resec-

tion of the pole of the kidney can also be applied in the management of diseases of dystopic kidneys with dyskinesia of the upper urinary tract and urinary stasis in the pelvis of the anomalous kidney.

On concluding this chapter we feel obliged to emphasize the clinical importance of this renal anomaly. We encounter it quite often in the clinic. We observed it in 1 per cent of patients with diseases of the kidneys and upper urinary tract. It is more often disclosed in children and at an age when a person is capable of work. Dystopic kidneys are predisposed to diseases such as hydronephrotic transformation, nephrolithiasis and pyelonephritis. These were encountered in 45 per cent of all cases. Besides, in 28.4 per cent there were manifestations of the anomaly itself, particularly pain which often caused patients with dystopic kidneys not complicated by disease to leave work. Thus, in 82.4 per cent of cases dystopic kidneys produced various clinical symptoms.

The causes of the special predisposition of dystopic kidneys to the development of various diseases stem from the defects of the anatomical structure of this anomalous organ. The flattened kidney with its unusual localization, in most cases unprotected by the ribs and often with peculiarities of the urinary tract (high and acutely angled origin of the ureter from the pelvis and its compression above the pole of the kidney, compression of the ureter by accessory vessels, often encountered with this anomaly, periureteritis, fixed kinking of the ureter, stricture of the ureter, frequently encountered extrarenal calyces), is conducive to the development of dyskinesia of the upper urinary tract and stasis of the urine in the renal pelvis. In addition, the dystopic kidneys exert mechanical pressure on the nerve ganglia and plexuses, vessels and surrounding organs, and cause a reflex effect on the function of various organs.

The clinical importance of this anomaly is also linked with the difficulties in its diagnosis. Patients with dystopic kidneys often receive unnecessary treatment for erroneously diagnosed diseases and tumours of the abdominal organs and undergo unwarranted operations (appendectomy, laparotomy). The diagnostic errors occur because diseases of a dystopic kidney have a clinical picture that is not typical of the organ: pain and tumour with unusual localization, dysuric and dyspeptic phenomena, abnormalities in the female genital sphere and nervous and vascular systems. Among these symptoms only dysuria and profuse haematuria, which is encountered very rarely, are indications of diseases of the urinary organ. In addition to these unusual symptoms of renal anomaly, those signs of renal diseases which develop are also atypical in these cases.

Urological examination is necessary when a tumour is found in the abdomen close to the vertebrae. Excretory urography and retrograde pyelography will show up the shape of the dystopic kidney and the position of the urinary tract characteristic of the anomaly, and also help to make the correct diagnosis of the anomaly and recognize the diseases of this kidney. The findings of tomography or pneumoretroperitoneum help to rule out fusion of the kidneys. Excretory urography, renography and scanning of the kidneys demonstrate renal function, while telepyeloscopy, cineurography or urokymography confirm or rule out dyskinesia of the urinary tract and urinary stasis in it. Renal angiography yields valuable information on vascularization of the kidney prior to surgery.

The peculiarities of the treatment of diseases of dystopic kidneys have an effect on the clinical course of this renal anomaly. Non-operative treatment in such cases brings only temporary relief. Pain leading to the loss of working ability and various diseases indicate the need for operation on dystopic kidneys uncomplicated by disease. Dyskinesia and urinary stasis in the pelves call for an expanded range of indications for surgery in many cases so as to prevent progression of the disease. In cases of pain caused by the dystopic kidney, nephrectomy is the most reliable operation. The results of nephropexy are often poor and call for repeated operations which are technically difficult because the surgeon has to manipulate on a kidney held in its seat by many vessels. Nephrectomy is indicated in diseases with marked morphological changes and non-functioning kidney and in cases of tumour. When renal function is maintained and dyskinesia and urinary stasis are found in the upper urinary tract, it is best to perform corrective operation with resection of the lower pole of the kidney and, whenever necessary, combine this with plastic surgery on the upper urinary tract or pyelolithotomy. Such operations improve the results of the surgical management of diseases of dystopic kidneys.

CHAPTER V

Anomalies in the Number of Kidneys

In our experience this large group of renal anomalies accounts for 49.6 per cent of all anomalies (696 persons).

Agenesis, aplasia, hypoplasia of the kidneys, and duplicated and supernumerary kidneys are included in this group.

AGENESIS AND APLASIA OF THE KIDNEYS

AETIOLOGY, PATHOGENESIS AND CLASSIFICATION

Renal agenesis is caused by the arrest of growth or anomalous vascularization on one or both sides at the stage of the development of the pronephros. In such cases half the urinary tract or, in bilateral affection the whole urinary tract, cannot develop.

The arrest of growth or disorders of vascularization in the pronephros stage may result in **aplasia of the kidney**. In such cases of disorders of the pronephros there are anomalies of the genitals and absence of a kidney, whereas in cases of a deficient mesonephric duct the ureter is absent, as a result of which an underdeveloped kidney may be found. Thus, depending on the stage of embryonal development during which the anomaly forms, different variants may be encountered. In earlier developmental stages various unfavourable factors induce agenesis of the kidney; in later stages aplasia of the organs may develop. These anomalies share clinical features in common and it is therefore useful to consider them together.

The first report of solitary kidney was made as early as 1543 by Vesalius (Sokolov, 1928), while bilateral agenesis was described in 1883 by Coen. Fedorov was the first to describe this anomaly in Russia in 1905. In later years quite a few reports have appeared in the literature based on occasional postmortem and clinical observations.

SURGICAL ANATOMY

In bilateral renal agenesis or aplasia both kidneys are absent and such foetuses are not viable. Unilateral agenesis or aplasia marked by the absence of one kidney is of importance in clinical practice.

MORPHOLOGICAL STRUCTURE

In agenesis or aplasia of one kidney the contralateral kidney is often hypertrophied. According to Sokolov (1928), a congenital solitary kidney was hypertrophied in 29 of 51 postmortem studies. In volume the solitary kidney is usually close to double the size of a normal kidney. The solitary kidney was found to be 16 to 18 cm long and weighed up to 440 g. The number of nephrons in a solitary kidney is equal to that in two normal kidneys (Maluf et al., 1957).

A congenital solitary kidney is often dystopic. Liokumovich (1911) and Rizvash (1935) each described such a case. In Sokolov's (1928) experience, in two of 51 cases the solitary kidney was dystopic.

The pelvis of a congenital solitary kidney is much larger than the pelvis of a kidney which has undergone compensatory hypertrophy at a late period in life. Its ureter may have various defects. Laughlin (1954) had a case of a retrocaval ureter of a solitary kidney, while Rivadeneyra (1968) had one of multiple stenosis of the ureter.

In aplasia the organ is very small and hardly resembles a kidney. It is usually a mass of fibrous tissue with small impregnations of dilated tubules scattered at random in the rich connective-tissue stroma; there are no glomeruli. Sometimes the kidney is lost in the mass of connective tissue and is a microscopical structure in which urinary tubules may be discerned only with scrupulous examination. In most cases such a kidney is not larger than 2 or 3 cm.

With this anomaly there is no renal pelvis and, at times, the ureter is also absent. In those cases when there is a ureter it varies in length; sometimes it is 1 or 2 cm long, or it may be of full length but terminates blindly at the pelvis. In some instances

the ureter ends blindly at the urinary bladder (Klosterhalfen, 1959) or has a narrow lumen (Crucioli et al., 1969). Nation (1944) described such a kidney with no ureter.

In some cases of renal agenesis or aplasia the whole trigone of the bladder or the corresponding half of it is absent. According to Nation (1944), the ureteral orifice on the side of the anomaly was absent in 11 of 16 cases. The ureteral orifice of the anomalous kidney sometimes drains into the genital tract and not into the bladder. According to Sokolov (1928), among 51 postmortem studies the ureter was absent in 12, was rudimentary in 2, had a blind end in another 2, and a cystic dilatation in 1.

In renal agenesis there is no vascular bundle. In some cases the aorta and vena cava inferior have small depressions corresponding to the renal vessels (Mkhitarov, 1930). In aplasia of the kidney rudimentary vessels in the form of thin branches may be encountered (Wermenski, 1969).

COMBINATION OF THIS ANOMALY WITH DEVELOPMENTAL ANOMALIES OF OTHER ORGANS

Aplasia and agenesis of the kidneys are often attended by various developmental anomalies of other organs, developmental anomalies of the female genitals in particular. This kidney anomaly has been found together with duplication of a bifid uterus with a septum and duplication of the vagina, one-horned rudimentary uterus; there have been cases of absence of the uterus, the vagina, the large and small pudendal lips, the clitoris, ovaries or seminal vesicles, testis; there have also been concomitant ureteral atresia, hypospadia or epispadia, atrophy of the prostatic lobe, hypoplasia of the testes, cryptorchism (Radice and Giordano, 1968).

The frequency of genital anomalies in renal aplasia is variable, according to the literature, and these combinations are more often found during postmortem examination than in patients in the clinic. Ballowitz (1895), for instance, among 213 studies of renal aplasia found 73 with anomalies of the genital organs, whereas Sokolov (1928) found only four out of 51.

Chawla et al. (1966) found five solitary kidneys among 11 cases of renal anomalies in 23 females who had no vagina.

According to Humphry and Munn (1966), a combination of anomalies of the kidneys and heart is a rarity.

STATISTICS

Aplasia and agenesis of the kidneys are relatively rare. By 1895 Ballowitz found information in the literature on 213 cases.

According to Gotlib, 453 cases had been described up to 1929, and only in six of them was a correct diagnosis made prior to surgery. In his research up to 1935, Savinykh found reports on 458 cases of such kidneys, Gimpelson found 700 up to 1939, and Klosterhalfen had already found a thousand by 1959. After that date we found information in the literature on another 182 cases, so that the sum total has now reached 1182.

Early reports dealt with one or two cases observed in a single clinic, whereas later some authors were able to report on eight to 16 clinical cases (Gimpelson, 1949; Lopatkin, 1961). We observed 18 patients with unilateral renal aplasia or agenesis in the clinic.

The anomaly prevailed on the right side (it was on the right side in 11 cases and on the left in seven). Sokolov (1928) and Gimpelson (1949) also noted a high incidence of the anomaly on the right side, although Nation (1944) claimed that it was encountered with equal frequency on both sides.

Bilateral renal agenesis is still rarer. According to Frumkin and Marz (1954), 170 cases of bilateral renal agenesis were described in the literature, including four cases from their own practice. Campbell (1966) considers it to be a very rare anomaly, being encountered in only 0.04 per cent of cases. The anomaly is not of clinical significance. It is usually found in non-viable foetuses.

Thus, the absolute incidence of renal aplasia and agenesis in 188 838 postmortem findings is 0.10 per cent (Table 5). Among

Table 5. Frequency of Renal Aplasia and Agenesis Discovered at Postmorten Examination

Author	Number of autopsies	Number of anomalies	%
Podobed (1912)	66 000	56	0.08
Sokolov (1928)	50 198	51	0.10
Campbell (1936)	26 480	40	0.15
Ageyenko (1938)	700	1	0.14
Nation (1944)	27 000	33	0.12
Smith and Orkin (1945)	18 460	13	0.07
Total	188 838	194	0.10

471 autopsies performed in cases of renal anomaly Smith and Orkin (1945) encountered 13 cases (2.76 per cent) with agenesis.

This anomaly accounts for 0.08 to 0.19 per cent of all diseases of the kidneys and upper urinary tract (Abeshouse, 1944). In our practice we observed it in 0.18 per cent of such cases, which

is in agreement with Abeshouse's data. This anomaly is therefore encountered more frequently in the clinic, which is evidently linked with the predisposition of a solitary kidney to pathological disorders and diseases.

Agenesis and aplasia of the kidneys are apparently encountered more frequently among men (Sokolov, 1928; Nation, 1944). Among our cases there were ten males and eight females. In Gimpelson's (1949) practice alone there was a slight preponderance of females.

The anomaly may be found in patients of any age. In our experience it was discovered at ages from five to 67 years (Table 6).

Table 6. Frequency of Renal Aplasia and Agenesis in Relation to That of Other Renal Anomalies

Author	Number of renal anomalies	Number of cases of aplasia or agenesis	%
Gimpelson (1949)	250	16	6.40
Pekarskaya (1955)	57	1	1.75
Denmukhamedov (1957)	205	2	0.97
Vourinen (1962)	69	2	3.03
Antonov (1964)	63	2	3.17
Rozhdestvensky (1965)	108	5	4.63
Plotnikov (1966)	71	4	5.63
Vapra and Tulmin (1967)	82	2	2.43
Total	905	34	3.75

According to the data in the literature, aplasia and agenesis accounted for 3.75 per cent of 905 kidney anomalies. Among our 1405 cases of various renal anomalies, this anomaly was found in 18, which accounted for 1.28 per cent of the total.

In agenesis or aplasia of one kidney the remaining kidney is evidently predisposed to disease. Evidence of this is the high incidence of the anomaly in clinical practice as compared to postmortem findings, as well as the high frequency of diseases of such kidneys found at postmortem examination. According to Savinykh (1935), Winter encountered disease in 38 per cent of solitary kidneys and Anders in 42 per cent. Sokolov (1928) found disease in 71 per cent of cases during postmortem examination. It is quite probable that a solitary kidney is more predisposed to disease because it functions continuously without rest. This cannot but intensify the wear of its glandular components.

Various diseases affecting a normal kidney have been encountered in solitary kidney, but pyelonephritis, stones, tuberculosis

and hydronephrosis are found most frequently (Gotlib and Frenkel, 1932; Sinkevichus, 1958). In our 18 clinical cases of a congenital solitary kidney, there were nine with affection of this organ (stones, pyelonephritis, glomerulonephritis, hydronephrosis).

The particular predisposition of this anomaly to disease calls for special preventive measures. It is obligatory that the people suffering such an anomaly be kept under strict medical control.

SYMPTOMATOLOGY, CLINICAL COURSE AND RECOGNITION OF RENAL AGENESIS AND APLASIA

A congenital solitary kidney uncomplicated by disease may produce no symptoms or only a very poor complex of symptoms, mainly a palpable enlarged kidney and moderate dull pain in the small of the back.

Only one of our three patients complained of pain. Thorough urological examination of all three failed to reveal any disease but one of these patients suffered dull pain of low intensity, though there was no referred pain typical of the anomaly. A palpable kidney called for urological examination. In this case we could not assuredly rule out the possibility of initial degenerative changes in the kidney. A five-year follow-up with regular examination, however, did not reveal any disease, which is why we have included this patient in the group with no diseases. It is possible that the dull pain was caused by moderate circulatory disorders in the hypertrophied kidney with no functional disturbances, the more so since the pain occurred only from time to time and was linked with a faulty diet.

A hypertrophied solitary kidney, particularly if it is dystopic, may be detected by palpation. We found this symptom in all patients of this group. A smooth elastic and enlarged kidney was felt in the hypochondrium, whereas in cases of dystopia it was palpated in the iliac region or in the pelvis.

According to the clinical course, the following three groups of aplasia and agenesis of the kidney can be distinguished: asymptomatic, with pathological disorders but without disease, and with disease.

Among 18 cases of this anomaly in our practice, there were four with an asymptomatic course. The anomaly was found by chance during a regular medical examination or during urological examination undertaken due to a recognized anomaly of the female genital organs.

R., a 14-year-old girl, was admitted to the clinic on January 18, 1961. She complained of acute pain experienced in the lower abdomen regularly

once a month. She said that she had been ill for a year and linked her disease with the first menstruation periods. Palpation above the pubis revealed a smooth mobile tumour which slid into the true pelvis. Digital rectal examination revealed an elastic, hard and painful infiltration shaped like a sausage ascending on the anterior surface of the rectum some distance from the anal opening. Haematometra in duplication of the genital tracts was suspected. Blood and urinaty tests showed normal values. In cystoscopy the orifice of the right ureter and this part of the trigone were absent. Excretory urography demonstrated the left pelvis and left ureter; on the right, the urinary tract was not visible. Pneumoretroperitoneum showed only the left kidney, the right kidney being absent. Aplasia of the right kidney and duplication of uterus and vagina were diagnosed. Laparotomy was performed. A duplicated uterus and a small amount of blood at the duplicated uteral adnexa were found in the abdominal cavity. The right uterus was continuous with an accessory vagina filled with blood. The vagina had a blind end. The uterus and accessory vagina were excised. The girl recovered.

In this case aplasia of the kidney was revealed due to an anomaly of the female genitals which called for urological examination. All symptoms were associated with diseases of the genitals.

As was mentioned above, in one of our cases the hypertrophied solitary kidney caused dull pain in the small of the back now and again. Dull pain of this localization and the palpable kidney could be mistaken for some disease of the kidney which, in turn, usually serves as an indication for a urological examination. Aplasia of the kidney is thus revealed and disease ruled out.

Renal agenesis and aplasia are clinically important because a solitary kidney cannot be removed in case of disease, while a diagnostic error may lead to such an operation. Reports in the literature on such fatal diagnostic errors are not rare. This anomaly acquires clinical significance also in cases in which a disease of the solitary kidney develops and it is necessary not only to diagnose it early, but also to treat it and determine the prognosis.

Aplasia and agenesis of the kidney may be suspected in healthy people when one enlarged kidney is palpated while the other kidney cannot be felt. This renal anomaly can also be suspected in a patient with anomalies of the genitals and also when renography shows that one of the kidneys is not functioning. Complete urological examination is necessary when aplasia or agenesis of the kidney is suspected.

The following methods of examination may be necessary in making a diagnosis of this anomaly: cystoscopy, excretory urography, retrograde pyelography, pneumoretroperitoneum, renal angiography and radioisotope study.

The diagnostic difficulties are usually associated with the differential diagnosis of a blocked kidney and this anomaly in which there is also no function of the kidney. The anomaly also has to be differentiated from some other renal anomalies such

as dystopic kidney, duplicated or unilateral fused kidney in ectopy of the ureteral orifice, and from some diseases producing a filling defect in radiocontrast studies.

In some cases of aplasia and agenesis of the kidney, cystoscopy reveals the absence of a ureteral orifice on one side and asymmetry of the trigone of the bladder; sometimes the trigone may be symmetrical but with a very small ureteral orifice. According to Zinser (1955), in one third of such cases there is no ureteral orifice in the bladder.

The presence of only one ureteral orifice in the bladder, however, is not an absolute sign of aplasia or agenesis, because the second orifice may sometimes open extravesically. In ectopy of one ureteral orifice, it is difficult to differentiate this renal anomaly from a solitary duplicated kidney, unilateral fused kidney or pelvic dystopia of the organ. Information from the medical history helps to determine the seepage of fluid by-passing the bladder, while X-ray examination helps to confirm the diagnosis of renal anomaly.

It should also be borne in mind that when there is only one ureteral orifice, the side on which it is found is not necessarily the side on which the kidney is located, because a solitary kidney may be crossed-dystopic.

When there are two ureteral orifices in the bladder and aplasia of the kidney, one of the orifices does not contract and indigo carmine does not flow from it.

Absence of the shadows of the kidney and pyelo-calyceal system on excretory urography on one side suggests a diagnosis of aplasia and agenesis of the kidney, particularly in patients in whom anomalies of the genitals have already been revealed.

N., an 18-year-old girl, was admitted to the clinic on April 26, 1963 for examination. Prior to this she had been in a gynaecological department where anomaly of the genitals was found (duplication of uterus and vagina). The right ureteral orifice in the bladder was not evident in chromocystoscopy; indigo carmine was not discharged on the right and appeared in three minutes on the left. The girl was sent to the urological clinic for further examination. With excretory urography (Fig. 87) the shadows of the left renal pelvis, calyces and ureter were seen on the radiographs. There were no shadows of the urinary tract on the right. Diagnostic pneumoretroperitoneum: the outlines of the left kidney were seen on the radiograph on the level of the 12th thoracic to the 3rd lumbar vertebrae. The outlines of the right kidney could not be seen. Diagnosis of aplasia of the right kidney was made.

When there is a ureteral orifice, a catheter can usually be advanced into it only for a short distance. With retrograde pyelography the ureter can be seen as a thread, short or of normal length, but terminating blindly in a narrowed end. In aplasia and agenesis of the kidney the pelvis is always absent. Because of the danger of infecting the solitary kidney, retrograde pyelo-

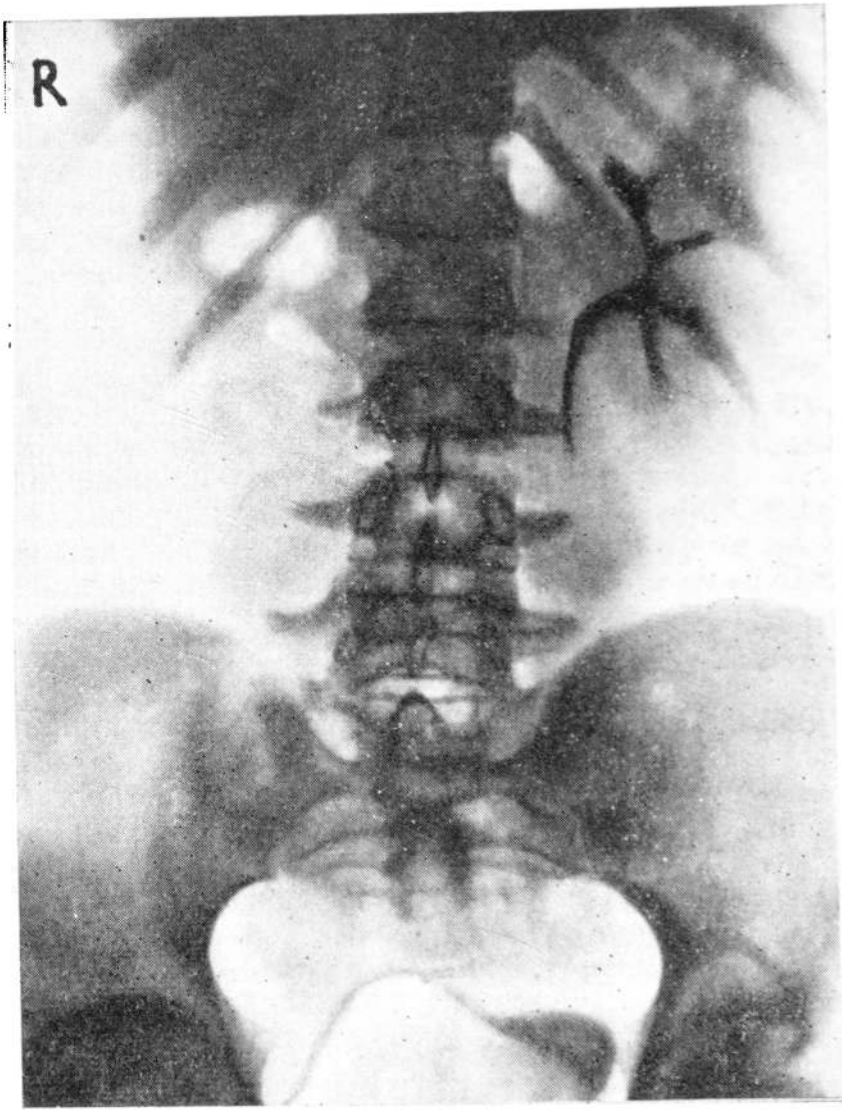


Fig. 87. Case N. Aplasia of the right kidney. Excretory urography

graphy should be conducted with great care and, whenever possible, avoided.

The diagnosis of this anomaly is particularly convincing when the radiographs produced in pneumoretroperitoneum show no outlines of one kidney, while the other kidney is large and uniformly hypertrophied. Pneumoretroperitoneum, however, is not always helpful in diagnosing the anomaly. The diagnosis cannot be made in cases of paranephritis and when the space free of the kidney is occupied by the spleen or liver. It is difficult to produce the image of the kidney in pelvic dystopia and when the kidney is small and has an ureter terminating blindly at

the bladder. In such cases renal angiography can eliminate any doubts in regard to the diagnosis.

Renal angiography is the most valuable method in the diagnosis of renal aplasia and agenesis, removing any doubt in difficult cases (Lopatkin, 1961). The absence of the renal artery on the side of the anomaly and the absence of the ureteral orifice in the bladder is the deciding factor. Angiograms in cases of aplasia and agenesis are identical. This method of examination also makes it possible to differentiate the anomaly from renal atrophy.

Scanning suggests aplasia if there is no image of the kidney. Brenez (1968) pointed out the value of this method in the diagnosis of such an anomaly.

PECULIARITIES OF THE SYMPTOMATOLOGY, CLINICAL COURSE AND RECOGNITION OF DISEASES OF A SOLITARY KIDNEY

In diseases it is usually the symptoms of the illness which reveal the solitary kidney.

Pain and tumour in cases of dystopic solitary kidney are sometimes localized in the abdominal cavity. Such a low localization may create difficulties in the diagnosis not only of the anomaly but also of the disease. In a case of unilateral renal colic, the patient may have anuria.

The course of a disease in aplasia of one kidney is characterized by the more rapid development of renal insufficiency. The diagnostic difficulties are caused not only by the need to recognize the disease, but to define the type of anomaly exactly and differentiate the symptoms of the anomaly and those of the disease by means of the methods of examination described above.

TREATMENT OF DISEASES OF A CONGENITAL SOLITARY KIDNEY

A solitary kidney does not usually require any treatment. Treatment is needed only when it is affected by disease. The occasional mention made in the literature of the removal of a solitary kidney (Shanatin, 1937; Ivashko, 1957) are examples of tragic errors which are usually fatal for the patient. The organ-preserving operations performed in cases of disease of a solitary kidney include pyelolithotomy, ureterolithotomy, nephropexy and plastic surgery on the urinary tract.

In cases of disease surgical intervention must be considered from the point of view of saving the solitary kidney. Operations have to be performed on such patients most frequently for stones of the kidney or ureter. We carried out operations on three pa-

tients. Different plastic operations have to be conducted in hydronephrosis of a solitary kidney, even plastic surgery of the urinary tract by means of intestinal grafts (Khomenko, 1963; Gaca and Wehinger, 1969) and nephropexy in severe nephroptosis.

RENAL HYPOPLASIA

AETIOLOGY, PATHOGENESIS, CLASSIFICATION AND SURGICAL ANATOMY

Hypoplasia of the kidney is the congenital reduction of its size. In this case the ureter is no different from a normal one and the contralateral kidney is not enlarged.

This anomaly results from impaired development of the parenchyma of the permanent kidney or its deficient vascularization.

Renal hypoplasia may be unilateral or bilateral. The degree of underdevelopment of the kidney varies from case to case. According to the experience of some authors, in a bilateral process the right kidney is usually altered more than the left.

MORPHOLOGICAL STRUCTURE

Hypoplastic kidneys vary in size usually from 2 to 5 cm, sometimes up to 7 cm; this is one third to one sixth the size of a normal kidney (Sokolov, 1928). In the newborn the hypoplastic kidney weighs 6-12 g (normal weight 40-50 g).

Rudimentary kidneys (very small kidneys with embryonic lobular structure and disproportionally small anteroposterior dimension, flattened, sometimes triangular) and dwarfed kidneys (resembling a normal kidney but small in size) are distinguished according to size. The larger the hypoplastic kidney, the more it resembles a normal kidney in shape and structure.

On section this anomalous kidney has cortical and medullary layers the boundaries between which may be erased to some degree. A kidney with a normal structure has a normal number of pyramids, while a considerably underdeveloped kidney is marked by morphological and quantitative anomaly of the nephrons (Habib et al., 1962).

Two types of renal hypoplasia are distinguished. One has a normal histological structure and the other is dysplastic with more or less excessively developed connective tissue, a small number of normal or rudimentary glomeruli scattered at random in the connective tissue, and thickened vessels and tubules often arranged in nest-like clusters. The glomerular capsule is considerably thickened. The connective tissue in a hypoplastic kidney is pierced by the bundles of smooth muscle fibres which are inher-

ent in an embryonic kidney (Poetzel, 1956). The formation of cartilages and even bones with bone marrow may be noted (Tedeschi and Holtham, 1952).

The rich content of the connective tissue in a hypoplastic kidney can be explained by the general law of biology, namely, that deficiently developed highly differentiated glandular tissue is replaced by connective tissue which does not usually shrink and does not develop into cicatricial tissue, as is the case with an inflammatory process.

As distinct from an atrophied kidney, a hypoplastic kidney has a smooth surface and an easily removable capsule. It contains no signs of interstitial nephritis. It should be borne in mind, however, that secondary inflammation may also occur in a hypoplastic kidney.

Because of the poor functioning of a hypoplastic kidney, the contralateral kidney is often hypertrophied, a fact noted by Sokolov (1928) in 23 of 50 cases.

In renal hypoplasia, the vessels are usually either normal or also hypoplastic: they are abnormally narrow, the walls of the arteries and veins are thickened, the artery of such a kidney is sometimes hypoplastic (Giombolini, 1968).

The pelvis of a hypoplastic kidney might be normal, reduced in proportion to the size of the kidney or, most frequently, even enlarged despite the absence of an obstacle in the ureter. According to Boissonnat (1962), three types of pelves of hypoplastic kidneys are distinguished. Most common is the upper polar type with the calyces arising from the upper pole of the pelvis. The bipolar type is encountered less frequently; the calyces in this case arise from the upper and lower poles of the pelvis. Finally, in the pseudo-miniature type small calyces are distributed along the entire periphery of the pelvis. In this case a high percentage of calyceal diverticula of unusual size and localization is noted.

The ureter of a hypoplastic kidney is often narrowed or obliterated along its entire length; its lumen may be evenly narrowed or sometimes dilated, but in some cases the ureter is absent. Tuchin (1972) and other authors noted cases with ectopia of the orifice of the ureter of a hypoplastic kidney into the genitals.

Dystopia of a hypoplastic kidney and dystopia of a duplicated hypoplastic kidney have been described (Topchibashev, 1957). In other reports, however, dystopia of the contralateral kidney is mentioned (Sucin and Caluser, 1966).

COMBINATION OF RENAL HYPOPLASIA WITH OTHER DEVELOPMENTAL ANOMALIES

As distinct from aplasia, hypoplasia of the kidney is rarely encountered in combination with developmental anomalies of other organs, including anomalies of the genital organs. This is associated with the fact that a hypoplastic kidney develops later in the embryonal period when the genital organs are already formed and develop further independently.

Occasional cases of hypoplastic kidney combined with epi- or hypospadias or pseudohermaphroditism can be found in the literature.

STATISTICS

Hypoplasia of the kidneys is quite rare. Gotlib found that up to 1929 62 such anomalies had been reported in the literature. Manuilov had found 73 reports by 1944. The actual number of such anomalies, however, is much greater, because some clinics to date describe three to seven cases each (Ritter and Krainer, 1950; Burkland, 1954). The Boeminhaus Clinic (1958) has information about 16 such cases. Bengtsson et al. (1970) presented data on 76 hypoplastic kidneys. In addition, individual cases are often reported in the literature.

Evidently, fewer cases of hypoplasia than of aplasia have been reported because of the lesser clinical importance of the former. We possess information on 38 patients with renal hypoplasia. Thus, including our own cases, we now have data on 430 cases of renal hypoplasia.

Bilateral renal hypoplasia is very rare. In unilateral hypoplasia, reports on the frequency of the side of the anomaly differ from author to author. Sokolov (1928) encountered twice as many cases on the right, while Gimpelson found twice as many cases on the left. Bengtsson et al. (1970) observed the anomaly with equal frequency on both sides. In our experience it slightly prevailed on the right (a total of 21 cases).

Table 7. Frequency of Renal Hypoplasia Found at Postmortem Examination

Author	Number of autopsies	Number of anomalies	%
Sokolov (1928)	50 198	50	0.09
Nation (1944)	27 000	45	0.16
Smith and Orkin (1945)	18 460	35	0.18
Total	95 658	130	0.14

It can be seen from Table 7 that the absolute frequency of renal hypoplasia on postmortem examination is 0.14 per cent. According to Campbell (1957), the anomaly is encountered in 0.2 per cent of patients on the average, among children in 0.17 and among adults in 0.21 per cent. Among 471 cases with renal anomalies Smith and Orkin (1945) found 35 cases of renal hypoplasia (7.4 per cent). Though cases of kidney hypoplasia are not described as frequently as aplasia in the literature, judging from the data its incidence is actually higher. The slightly higher frequency of the anomaly among adults may be linked with the fact that in some cases it was mistaken for renal atrophy.

Among cases of diseases of the kidneys and upper urinary tract, hypoplasia of the kidney was noted in 0.08 to 0.56 per cent (Dees, 1941; Rozhdestvensky, 1965). In our practice it was found in 0.58 per cent of such patients. The higher frequency of renal hypoplasia in clinical practice is obviously linked with the diseases it has induced.

Hypoplastic kidneys are found more frequently in females (Gimpelson, 1949). According to the postmortem data of Sokolov (1928), hypoplasia of the kidney was encountered in females in 0.13 per cent of cases, and in males in 0.01 per cent. In our experience there were 23 females and 15 males.

This anomaly is found in people of any age. We had patients from eight to sixty-two years old, but most of them were in the 39-49 age brackets, which may be linked with the increased load experienced at this time of life.

Judging from Table 8, renal hypoplasia accounts for 5.0 per cent of all renal anomalies. This is more than aplasia and agenesis of the kidneys account for. Among 1405 different renal anomalies we had 38 cases of renal hypoplasia (2.7 per cent).

Hypoplastic kidneys are a frequent cause of arterial hypertension (Higbel, 1944). Severe arterial hypertension was noted in

Table 8. Frequency of Renal Hypoplasia Among Other Kidney Anomalies

Author	Number of renal anomalies	Number of cases with kidney hypoplasia	%
Dees (1941)	135	8	5.8
Gimpelson (1949)	250	13	5.2
Smith and Orkin (1945)	471	35	7.4
Pekarskaya (1955)	57	2	3.5
Antonov (1964)	63	3	4.6
Rozhdestvensky (1965)	108	5	4.6
Vapra and Tulmin (1967)	82	3	3.6
Total	1166	59	5.0

half of patients with bilateral renal hypoplasia (Habib et al., 1962).

Different diseases are often found with this anomaly: chronic pyelonephritis, glomerulonephritis, hydronephrosis, nephrolithiasis, nephroptosis. These may, in turn, be the cause of arterial hypertension. In our practice there were 17 cases of diseases of a hypoplastic kidney: pyelonephritis, 3; nephrolithiasis, 7; glomerulonephritis, 3; hydronephrosis, 4. In six cases the hypoplastic kidney was the cause of severe arterial hypertension.

The high frequency of diseases and abnormalities in patients with this anomaly calls for regular medical examination and, in many cases, for treatment.

SYMPTOMATOLOGY, CLINICAL COURSE AND RECOGNITION OF HYPOPLASTIC KIDNEY

A hypoplastic kidney may produce no symptoms, but in some patients an anomalous kidney with no marked disease causes clinical symptoms among which pain and arterial hypertension should be emphasized.

According to Emmett and Alvarez-Ierena (1952), lumbar pain was noted in half of 68 patients with hypoplastic kidneys. In some of these cases the pain could hardly be attributed to the kidney because it was felt in the abdomen. Campbell (1957) links the pain in such cases with sclerotic compression of the nerve endings. The same mechanism promotes the later development of arterial hypertension.

Arterial hypertension often attends renal hypoplasia. Ekstrom (1955) found it in twenty of 179 patients with hypoplastic kidney; Emmett and Alvarez-Ierena (1952) diagnosed arterial hypertension in 37 per cent of 68 patients with removed hypoplastic kidneys in which no disease had been revealed by histological examination. At the same time, the percentage of contracted kidneys among cases of pyelonephritis in the practice of these authors was lower. It is possible that the cause of arterial hypertension in people with kidney hypoplasia is sclerotic compression of the anomalous vessels of the kidneys. Pytel (1960) thinks that arterial hypertension in such cases is caused by secondary pyelonephritis.

Other clinical symptoms in renal hypoplasia are very rare. According to Poetzel (1956), urine excretion is sometimes diminished. Szibert (1956) observed in one of his patients with hypoplastic kidney profuse haematuria, which he attributed to rupture of the anomalous vessel of such a kidney as the result of disorders in its dynamics.

Poor growth, low weight and renal osteodystrophy are signs of bilateral renal hypoplasia in infants (Habib et al., 1962).

In our group of patients, 21 had hypoplastic kidneys uncomplicated by disease. Sixteen of them had clinical symptoms: pain in ten cases and arterial hypertension in six. The other patients did not have any clinical symptoms of the renal anomaly or its diseases.

It is interesting that in four of our six patients arterial hypertension developed after pain.

Pain in hypoplasia of the kidneys was felt in the lumbar region and rarely in the abdomen. It was dull in character and usually occurred in periods, although in some cases it was continuous. Arterial pressure in six patients ranged from 155/80 to 210/100 mm Hg.

According to the clinical course of renal hypoplasia, three groups of patients can be distinguished: without symptoms, with pathological phenomena, with disease. We encountered an asymptomatic course in five of 21 cases. The hypoplastic kidney was discovered by chance during examination for diseases of other organs or of the contralateral kidney.

G., a 42-year-old male, was admitted to the clinic on February 19, 1965 complaining of dull pain and tumour in the left hypochondrium. He had been ill for three years. An enlarged, painless and smooth structure was palpated in the left hypochondrium with ballottement of the lumbar region. Pasternatskii's sign was negative. Blood and urine tests showed no abnormalities. With excretory urography the shadow of the contrast medium was seen on the left in the form of round spots, on the right the shadow of a very small pelvis with proportionally smaller calyces was seen against the background of regular outlines of a small elongated kidney. A diagnosis of hydronephrosis of the left kidney and hypoplasia of the right kidney was made. Operation was performed. The following diagnosis was made from inspection: hydronephrosis induced by compression of the ureteropelvic segment by an accessory vessel. Plastic surgery was undertaken with excision of the stenosed ureter with part of the renal pelvis. Sutures were applied to the pelvis and the ureter was stitched into its lower corner. The patient recovered.

In this patient, hydronephrosis of the contralateral kidney served as the motive for urological examination. Because of hypoplasia of the other kidney, plastic surgery was necessary despite the diminished function of the hydronephrotic kidney.

In 16 of 21 patients of this group pain or arterial hypertension was the cause for urological examination, during which the hypoplastic kidney was discovered. These patients had complained for several years of dull pain in the small of the back or in the abdomen, which either appeared periodically or was continuous. Some of them developed arterial hypertension later. In some cases these symptoms were noted from childhood.

D., a 15-year-old girl, was admitted to the clinic on June 3, 1963 complaining of dull pain in the small of the back and high arterial pressure. She had experienced the dull lumbar pain from childhood. During the last two years

increased arterial pressure was found during prophylactic examination. The kidneys were not palpable. Pasternatskii's sign was negative. Arterial pressure, 165/85 mm Hg; blood and urine tests showed normal values. During excretory urography the shadow of a normal left renal pelvis was clearly demonstrated, but the right pelvis was small with miniature calyces. Pneumoretroperitoneum demonstrated regular outlines of a small right kidney and normal outlines of the left kidney. Diagnosis of hypoplasia of the right kidney was made. Nephrectomy was performed. Histological examination of the removed small kidney with a smooth surface showed a small amount of normal glomeruli, proliferation of connective tissue and bundles of smooth-muscle fibres. The diagnosis of kidney hypoplasia was confirmed. The postoperative period was uneventful. Arterial pressure dropped to normal level (125/65 mm Hg). Pain was completely relieved.

Bilateral hypoplasia of the kidney is manifested clinically between the ages of one and eleven years. In such cases the course is often complicated by renal insufficiency, which depends on the degree of congenital defects in the renal parenchyma and on complications, usually resulting from infection.

The diagnosis of hypoplasia of the kidney is often very difficult. Emmett and Alvarez-Ierena (1952) claim that in 30 per cent of cases the function of the hypoplastic kidney is maintained. In cases with preserved function, chromocystoscopy is of no value for the diagnosis because it offers no evidence of the anomaly. When the hypoplastic kidney functions poorly it helps to reveal the anomalous organ and arouses one's suspicions of its disease. In such cases X-ray examination becomes essential, during which the anomalous kidney will be confirmed.

A hypoplastic kidney is suspected when, with a disease in one kidney, symptoms of renal insufficiency occur.

The absence of the function of a kidney or its diminution may be revealed by isotope examination. The diagnosis of the anomaly is then made during subsequent X-ray examination.

The diagnostic difficulties are linked with the need to distinguish a hypoplastic kidney from a kidney that has atrophied through inflammatory degeneration processes. The findings of histological examination may be of assistance in this regard. In kidney hypoplasia the cortical and medullary layers are not differentiated, or else the kidney has a normal structure with mild cicatricial and inflammatory processes. The presence of the anomaly cannot be doubted in cases when a very small kidney with a normal histological structure is found.

X-ray examination is the principal method used in the diagnosis of renal hypoplasia. In some cases the outlines of the small kidney may even be seen on a scout radiograph. In such cases, however, the diagnosis has to be verified by other methods of examination.

Excretory urography often makes it possible to recognize the anomaly when the function of the kidney is maintained. In

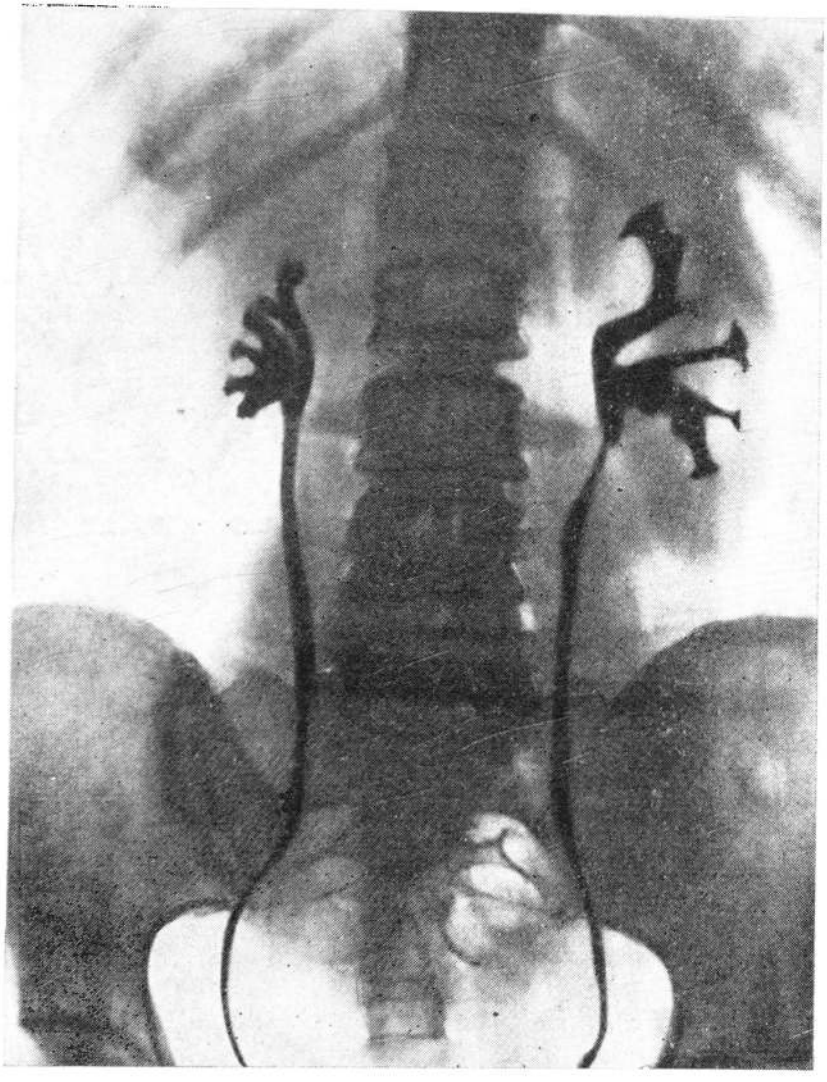


Fig. 88. Case N. Hypoplasia of the right kidney. Retrograde pyelography

these cases the radiographs show a small triangular or ampullary pelvis with small calyces against the background of a small shadow of the kidney or a miniature pelvis and calyces with all the X-ray signs of a normal kidney.

When a hypoplastic kidney functions poorly, there is no shadow of the pyelo-calyceal system, which permits us to presume a disease of the kidney and undertake additional examination.

The absence of an image of the pyelo-calyceal system on excretory urography obliges the examiner to resort to retrograde pyelography. The pyelogram in renal hypoplasia has the following characteristic signs: very small pelvis and normally shaped calyces; a small ampullary pelvis without calyces; absence of

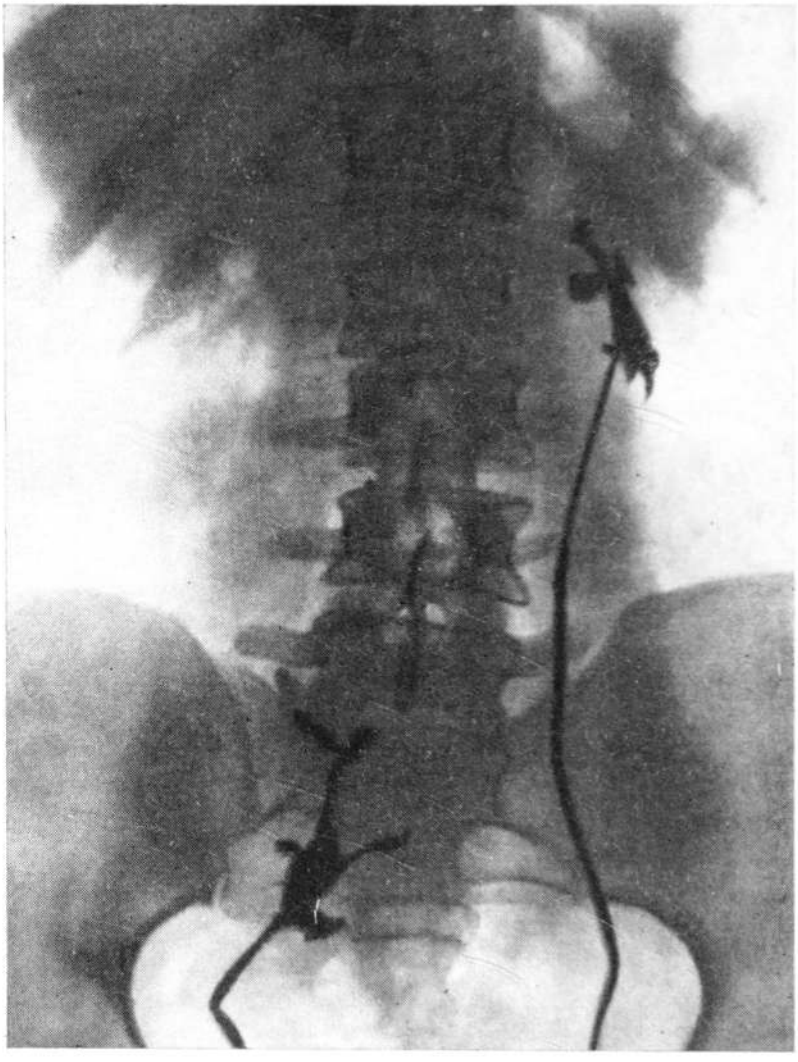


Fig. 89. Case Ts. Hypoplasia and lumbar dystopia of the left kidney, iliac dystopia of the right kidney. Retrograde pyelography

the major calyces with the minor calyces draining directly into the pelvis; a small rudimentary pelvis with one or two calyces as in an accessory pelvis of a duplicated kidney; a small rudimentary pelvis with the calyces directed upward or toward the vertebrae. In such a case the pelvis of the anomalous kidney is often dilated. Pyelograms which show a small shadow of the anomalous kidney are particularly demonstrative (Figs. 88 and 89).

We had an interesting case of hypoplasia of one kidney in bilateral renal dystopia. It is thus apparent that there can be a combination of signs of several renal anomalies in the same patient.

Pneumoretroperitoneum helps in the diagnosis of renal hypoplasia in doubtful cases. This method, particularly if combined with excretory urography or retrograde pyelography, usually shows a clear image of a small kidney with regular outlines. Inflammatory changes in the kidney are often combined with a sclerotic process in the perirenal fat, in which case no image of the kidney is produced on the radiograph. This sign is of some differential importance in distinguishing hypoplasia from atrophy of the kidney.

With inadequate preparation of the patient for examination the renal pyelo-calyceal system and the kidney itself can be clearly revealed by combining excretory urography with pneumoretroperitoneum and tomography.

Renal angiography is very important in the diagnosis of a hypoplastic kidney. Lopatkin (1961) claims that it makes possible a certain differentiation of hypoplasia from atrophy of the kidney. In hypoplasia the extra- and intrarenal vessels are uniformly reduced in calibre, whereas in atrophy the calibre of the intrarenal vessels is reduced or they are obliterated but the lumen of the renal artery is normal.

It is our experience that in renal hypoplasia urokymography reveals diminished contractions of the upper urinary tract.

Scanning is extremely valuable in the diagnosis of renal hypoplasia and its differentiation from atrophy. The scans show a functioning hypoplastic kidney as a very small organ. In a nonfunctioning hypoplastic kidney and in atrophy of the kidney no image is produced.

The anomaly possesses independent clinical significance because it may have clinical manifestations and may be recognized from the data of the examinations discussed above. It will be found far more frequently when the possibility of its existence is kept in mind. A very small hypoplastic kidney does not differ from aplasia of the kidney in clinical significance.

It is essential to recognize this anomaly before surgical intervention on the other kidney so as to prevent removal of the only functioning organ.

PECULIARITIES OF THE SYMPTOMATOLOGY, CLINICAL COURSE, AND RECOGNITION OF DISEASES OF A HYPOPLASTIC KIDNEY

The clinical picture of affections of a hypoplastic kidney is usually characterized by symptoms of the disease and does not differ from that of diseases of a normal kidney.

It needs only be noted that pyelonephritis of a hypoplastic kidney often induces the development of high and persistent

arterial hypertension. Such patients often die from myocardial infarction or uraemia.

With the development of a disease of a hypoplastic kidney, the X-ray picture usually loses the signs of hypoplasia and in such cases an erroneous diagnosis of renal atrophy consequent upon some disease is often made. The symptoms here are the same as those in diseases of normal kidneys: a filling defect in tumours, the' charred tree' symptom in pyelonephritis, etc.

TREATMENT OF HYPOPLASIA OF THE KIDNEY AND ITS DISEASES

Nephrectomy is the method of treatment when high arterial hypertension develops in a patient with renal hypoplasia. In Higbee's (1944) experience, the high arterial pressure dropped within a period of two years after this operation.

Among our patients five underwent nephrectomy for the management of arterial hypertension. Four of them were cured. The following case is given in illustration.

Z., a 19-year-old boy, was admitted to the clinic on November 19, 1969. He complained of rare episodes of dull pain in the right lumbar region and high arterial pressure. He had been ill for three years. The kidneys were not palpable. Pasternatskii's sign was negative. Arterial pressure 170/90 mm Hg. Blood and urine tests were normal. A scout radiograph showed no abnormalities. During excretory urography the left pelvis was clearly seen on a level with the first lumbar vertebra; the right very small pelvis had calyces directed upward. The findings of pneumoretroperitoneum confirmed the diagnosis of hypoplasia of the right kidney. Right nephrectomy was carried out. After the operation, arterial pressure decreased to 125/70 mm Hg. Two months later pressure again rose to 140/80 mm Hg, but then returned to normal levels and remained thus for a follow-up period of three years.

Histological examination of the removed kidney showed normal histological structure, thickened arterial walls, and smooth-muscle fibres among the fibres of the connective tissue. The diagnosis of hypoplastic kidney was confirmed.

This operation had good results in patients with hypertension of short duration (up to three or four years). In stable and long-term arterial hypertension nephrectomy is ineffective.

Pain associated with the hypoplastic kidney, sometimes quite intense, indicated the need for nephrectomy in three of our cases.

Non-operative treatment of diseases of a hypoplastic kidney does not differ from that applied in lesions of normal kidneys. An inflammatory process in bilateral renal hypoplasia responds well to treatment in patients under 12 years of age. In older patients an increased content of urea in the blood serum is very often found with subsequent development of azotaemia. Such

patients die in two to eight years, on the average, from renal insufficiency. Only a few live to the age of twenty.

Removal of the anomalous kidney is indicated in chronic pyelonephritis with high arterial hypertension when the process is unilateral. Our patients with this condition recovered after nephrectomy.

In other unilateral diseases of a hypoplastic kidney (stones, tumour, hydronephrosis, tuberculosis, etc.) and adequately functioning contralateral kidney, nephrectomy is also indicated. The operation is technically simple in such cases and involves the removal of a functionally inadequate kidney. Conservative operations are admissible in a well-functioning dwarfed kidney, but such conditions are rarely encountered. Gimpelson (1949) recommended nephropexy in cases of nephroptosis. The value of such an operation seems doubtful to us.

Nephrectomy is the operation of choice in this renal anomaly. We performed 23 such operations: 9 in hypoplasia with arterial hypertension or pain, and 14 with different diseases (three cases of pyelonephritis, four of hydronephrosis and seven of nephrolithiasis).

Diseases of the contralateral kidney in renal hypoplasia can be managed only by organ-preserving operations. The remaining hypoplastic kidney, less than one third the normal size, cannot provide for the organism's viability. One cannot hope for compensatory hypertrophy of such an anomalous kidney in these cases. Following removal of the contralateral kidney, death occurs from progressive renal insufficiency.

DUPLICATED KIDNEYS

AETIOLOGY AND PATHOGENESIS

There are several theories on **kidney duplication.** Gill (1952) believes that during the formation of the permanent kidney an accessory bud appears in the wall of the ureteral bud, already formed and actively growing in the metanephrogenous tissue. This accessory bud gives rise to still another ureteral branch. Both ureteral branches grow into the metanephrogenic tissue and form separate pelves and a split ureter which opens into the urinary bladder by means of a single orifice. Depending on the level of the accessory ureteral bud, the site of ureteral splitting may be at various distances from the pelvis.

According to Bremer (1957), a duplicated kidney with complete duplication of the ureter forms when an accessory ureteral bud appears in the caudal segment of the Wolffian duct, both

ureters then growing simultaneously. As in the case described above, both ureters grow into the metanephrogenic tissue and form two separate pelves, but they open into the urinary bladder by means of two orifices. Baumbush (1957) allows that such ureters may form from separate buds, but he does not rule out the possibility of their forming as the result of splitting of the primary ureteral bud.

The absence of a lumen in the initial parts of the tubular protrusion in the wall of the Wolffian duct, which gives rise to the ureter, results in the formation of a ureter with a blind terminal segment. In this case there is no ureteral orifice in the bladder. A ureter whose cranial segment terminates blindly is formed in the arrested development of one or two adjacent ureters or of one of the branches of the split ureter before it grows into the metanephrogenic tissue (Bremer, 1957).

Chwalla (1927) gave an interesting explanation of the development of ectopia of the ureteral orifice. He claimed that at a definite stage of embryogenesis the Wolffian ducts and the main and accessory ureters arising from them drain by means of a common passage into the cloaca. As soon as the bladder begins to form, the Wolffian ducts and the ureters separate. As a result, the ureters are displaced to the posterior wall of the bladder, while the Wolffian ducts and Mueller's duct stretching between them are displaced caudally into the formed urogenital sinus. Since the accessory ureter is cranial to the main one and originates somewhat later, its orifice may be displaced into the urogenital sinus in delayed separation from the Wolffian duct. Delayed separation of the ureter and its fusion with Mueller's canal is possible. In female-type formation of the foetus Mueller's canal is transformed into the uterus and vagina and the ureter is displaced into these parts of the genitals. In the male type of foetal development the ureter opens into the prostatic segment of the urethra which develops from the urogenital sinus or into the ejaculatory or deferent ducts developing from the Wolffian duct. Meyer (1907) and Baranov (1911) believed that ectopia of the ureteral orifice occurs because of the high position of the ureteral bud in the Wolffian duct. The lower end of the Wolffian duct is displaced into the bladder and becomes part of it, together with the ureteral orifice forming the trigone of the bladder. If the distance between the cranial and caudal ureters is short, both orifices have time to move into the bladder. When this distance between the ureteral buds is greater, the bud which is lying cranially does not have time to be displaced into the bladder before the Wolffian duct differentiates into the cystic and sinus parts, and so opens into the organs formed by the urogenital sinus. According to this theory, therefore, the extent of dis-

placement of the accessory ureter is determined by its level in the Wolffian duct and not by the time of its separation.

The bud of the main ureter is situated at a lower level in the Wolffian duct and because of this its orifice in the bladder is usually located higher than and lateral to the accessory ureter, which, together with part of the Wolffian duct, is displaced downward and medially. The lower intersection of duplicated ureters at the bladder is explained by this about-turn of 180 degrees of the Wolffian duct during its development. The upper intersection of the ureters apparently forms as the result of the rotation of the kidney and its migration to the lumbar region.

CLASSIFICATION

A duplicated kidney is a somewhat enlarged kidney with two separate and non-communicating pelves and two ureters. A contralateral kidney is also present (Fedorov, 1923). This definition characterizes the anomaly in full.

If we exclude the existence of a contralateral kidney, as suggested by Ishchenko (1927), it will be difficult to distinguish a duplicated kidney from a congenital solitary kidney and fused kidneys, which are independent developmental anomalies.

Other designations of a duplicated kidney can be found in the literature: accessory (Parmenov, 1941), separate (Petten, 1959), supernumerary (Teplitsky, 1935), accessory and pseudo-duplicated (Papin and Eisendrath, 1927), double (Vvedensky, 1931), hyperplastic with two pelves, etc. Parin (1929) designated a duplicated kidney double kidney, but in addition he also distinguished an accessory kidney, the smaller half of which is separated by a groove.

On the basis of the findings of angiography, Lopatkin (1961) considers a duplicated kidney to be one with a clear boundary between separately vascularized parenchymal areas. In addition, he distinguishes duplication of the renal parenchyma and duplication of the renal pelves. This distinction, however, can be made absolutely only by means of renal angiography, which is rarely called for in this developmental anomaly. It is for this reason that for clinical purposes it is recommended that any kidney with two pelves be considered as duplicated.

In almost all classifications a duplicated kidney is regarded as a renal anomaly and separately in the group of ureteral anomalies (Papin and Eisendrath, 1927; Zakharyan, 1941). In Gimpelson's classification (1949), the duplicated kidney is referred to as an anomaly of number. One must agree with his assertion that there cannot be an anomaly of the renal pelves without a renal anomaly, and vice versa. For this reason there are no grounds

for excluding this anomaly from the group of ureteral anomalies.

Duplicated kidneys are often accompanied with congenital ureteral defects which are very important in clinical practice, which is why we consider Kalenkovich's suggestion (1970) useful. He proposed distinguishing not only complete and incomplete duplication of the kidneys, i.e. conditions where there are two independent ureters or the splitting of the ureter of a duplicated kidney, but also subgroups with and without congenital ureteral defects in each group.

Ectopia of the orifice of the main or accessory ureter, ureterocele, blind ending of the terminal or cranial ureteral segment, and stenosis of the ureter or its valves are among congenital ureteral defects.

SURGICAL ANATOMY

A duplicated kidney is usually larger than a normal kidney and weighs more, but it can be the size of a normal kidney or smaller (Marochkin, 1942). In most cases it is bean-shaped and, more rarely, spindle-shaped. The upper part of such a kidney usually constitutes 15 to 30 per cent of the whole organ, but at times it may be larger than the lower part. According to Petten (1959), the seeming enlargement of the organ does not affect the number of pyramids because there are as many of them as in a normal kidney. The surface of a duplicated kidney is smooth, although in some cases the embryonic lobular structure is retained. Frequently a groove is seen on the surface of the organ as if separating it into two parts. The groove is not always clearly defined and does not always separate the organ fully. The renal capsule in this area is removed with difficulty. Some authors believe that in only 15 per cent of cases is the kidney completely separated into two parts by means of a connective-tissue layer. This borderline area consists of coarse connective tissue with impregnations of distorted tubules and hyalinosis of vessels (Ediny, 1953).

Neither part of the duplicated kidney has any peculiarities of histological structure. According to Ermolin (1964), however, the upper part contains less normal renal tissue and underdevelopment of the parenchyma is more marked.

A duplicated kidney is situated in the lumbar region at the level of the 12th thoracic and the 3rd or 4th lumbar vertebrae.

The urinary tracts of a duplicated kidney consist of several pyelo-calyceal systems and ureters arising from them, in most cases made up of two pelves and the ureter of the duplicated kidney. However, three, four and even five pelves and ureters may be encountered in a single such kidney.

The pelves of this anomalous kidney are usually arranged one under the other at some distance. The upper pelves are usually smaller than the lower ones (sometimes they are very small) and have one, two, or sometimes three calyces which are often intrarenal. The lower pelvis of a duplicated kidney is no different from normal, except that its upper calyx is often flattened or displaced downward, corresponding to the groove separating the parts of the organ. This sometimes creates difficulties in differential diagnosis with tumour of the kidney.

In the case of this renal anomaly the ureters may stretch separately from each other along their entire length and open into the bladder, each on its own side, by means of separate orifices. This is a case of complete duplication of the kidney and ureters. When the ureters merge along their length and terminate in the bladder by means of a common orifice, this constitutes incomplete duplication of the ureters or splitting. Ureteral splitting may occur at any level, but most frequently its site corresponds to the boundary of the separate cystoids. The ureters usually stretch in loose connective tissue, surrounded by a common connective-tissue sheath. These ureters are usually of the same calibre but in some cases that of the upper pelvis is narrower.

As early as 1887 Weigert discovered that the ureter of the upper pelvis of a duplicated kidney drains into the bladder caudally to the orifice of the second ureter. Meyer (1907) added to this that the medial orifice belongs to the ureter of the upper pelvis. These regular features are known in the literature as the Weigert-Meyer law, according to which the lower and medial orifices correspond to the upper pelvis. Both ureters often intersect, at times twice, as they stretch between the orifice and the pelvis. This law may be applied in cases of total duplication of the ureters. Judging from the literature and our observations, exceptions to the law are rare.

Duplicated kidneys can be uni- or bilateral. The following variants are encountered: unilateral duplicated kidney with complete duplication of the ureters; unilateral duplicated kidney with splitting of the ureter; bilateral duplicated kidneys with complete duplication of the ureters; bilateral duplicated kidneys with splitting of the ureters; bilateral duplicated kidneys with complete duplication of the ureters on one side and splitting of the ureter on the other.

Apart from these, there are also other variants apparently associated with later defects in the development of the different types of ureters. Blind termination of one of the ureters at the pelvis, noted in some of our cases, deserves special attention. In this case the upper part of the kidney, where the ureter ter-

minates blindly, consists of an area of cicatricial tissue with impregnations of deficiently developed tubules, which is usually observed in renal aplasia. The ureter may terminate blindly at different levels. In the literature it is sometimes called diverticulum of the urinary bladder.

The ureter of a duplicated kidney may terminate blindly at the bladder, while the lumen of its higher segment is preserved. Cases are described in the literature with the ureter of a duplicated kidney ending blindly at the bladder; in some the blind end of the ureter was lost in the tissues at the spine or terminated in the submucous layer of the bladder as a flask-like dilation (Karafa-Korbut, 1908; Schultze, 1967). Among our patients there were some in whom the accessory ureter of the duplicated kidney terminated blindly in the inguinal region or close to the bladder. These malformations are usually encountered in the accessory ureter, but there are descriptions of their being found in the main ureter of the duplicated kidney (Dubovik et al., 1967).

The orifice of one of the ureters of a duplicated kidney can often be found beyond the trigone of the bladder, whereas in bilateral duplication one orifice on each side may be so misplaced. In most cases this applies to the orifice of the accessory ureter, but ectopia of the main, or in ureteral splitting of the common, orifice cannot be ruled out. Such an unusual orifice lies in the diverticulum or neck of the bladder, in the urethra, at the entrance to the vagina or in the uterus in females or in males in the seminal vesicle, the ejaculation duct, etc.

Ureterocele is a frequent occurrence in duplicated kidneys; it is usually encountered in the accessory ureter and may also be bilateral. Vascularization of the duplicated kidney may be provided by one or more renal arteries, which separate into branches at the organ and give off separate terminal arteries to each of its parts. Each segment of such a kidney may have several arteries (Lopatkin, 1961). The veins are located correspondingly to the arteries. The lymphatics are also separate for each part of the duplicated kidney. The innervation of this anomalous kidney has no special features.

Duplication of the urinary tract is often encountered in other renal anomalies, i.e. in hypoplasia, fused kidneys and dystopic kidneys. Descriptions may be encountered of a combination in one kidney of elements of urinary-tract duplication, hypoplasia and dystopia, as well as hypoplasia, polycystic kidney and duplication of the urinary tract. Various renal anomalies may also be found in the same patient. There may be renal hypoplasia on one side and duplication of the kidney on the other, or dystopia of one kidney and duplication of the other.

COMBINATION OF A DUPLICATED KIDNEY WITH DEVELOPMENTAL ANOMALIES OF OTHER ORGANS

There are reports in the literature of the combination of a duplicated kidney with a bilocular urinary bladder (Wehrbein, 1940), diverticulum of the bladder and multiple defects in the urinary system (Payne, 1959).

Since the urinary and genital organs form from common buds, a combination of anomalies of these two systems frequently occurs. Hypospadia and other developmental anomalies of the external and internal genitals may be encountered in renal duplication (Lefebvre et al., 1966).

Combination of a duplicated kidney with anomalies of more remote organs are a less frequent occurrence. It may be found in some developmental anomalies of the bone system (sacralization, spina bifida, duplication of the rib, etc.), cleft upper lip and bilateral clubfoot (Novikova and Gotovets, 1967), hypoplasia of the viscera (Volpyan, 1964), duplicated gallbladder (Mastika, 1967), trilocular spleen (Krestovsky, 1939) and the Shereshevsky-Turner syndrome (Tikhonov, 1968).

STATISTICS

Duplicated kidneys occur quite frequently. Their absolute frequency, according to postmortem findings, ranges from 0.5 to 1.1 per cent (Campbell, 1936; Zschoch, 1962). The anomaly was found in 0.5 to 4.8 per cent of all patients at one urological clinic (Smith and Orkin, 1945; Kohnlein, 1960).

Duplicated kidneys are found even more frequently among patients with diseases of the kidneys and upper urinary tract. The anomaly was revealed in 638 of our patients, which accounted for 3.4 per cent. Operations on a duplicated kidney accounted for 4.7 per cent of all operations on the kidneys and upper urinary tract in our practice. According to Fedorov (1923) and Vvedensky and Stekolnikov (1934), operations on duplicated kidneys constituted 0.5 to 3.3 per cent of all operations on kidneys and the upper urinary tract. The somewhat higher percentage in our experience can perhaps be explained by the fact that the anomaly is revealed more frequently at our clinic.

Among patients examined by excretory urography at our clinic, 1.6 per cent were found to have duplicated kidneys (Kalenkovich, 1970). According to other authors, this percentage is still higher and ranges from 1.7 to 3.0 per cent (Rayne, 1959; Dhawan et al., 1965). Cystoscopy reveals duplicated kidneys in 0.3 to 1.5 per cent of all patients examined (Arsh, 1932; Filippova, 1940).

Duplicated kidneys are apparently encountered more frequently among females. In our experience they accounted for 77.5 per

cent of cases. This was emphasized by Nation (1944) and Plotnikov (1966). The anomaly is observed in patients of any age, though it is revealed more frequently up to 40 years of age. Gimpelson (1949) noted this also among his cases. Duplicated kidneys are described particularly often in children (Tudor et al., 1962; Novikova and Gotovets, 1967), in 3 to 17 per cent of cases. It can be presumed that the more frequent recognition of patients with duplicated kidneys at a younger age is linked with the clinical manifestations of the anomaly and of diseases of the anomalous kidney, which call for examination and identification.

Duplicated kidney rates first among renal anomalies in frequency. We encountered this developmental anomaly in 638 of 1405 renal anomalies, that is, in 45.5 per cent. In the experience of Dees (1941) and Ferber (1963) duplicated kidneys accounted for 29 to 55.8 per cent of all renal anomalies.

Unilateral duplication of kidneys is encountered most often, being found in 80.3 per cent of our patients, while only 19.7 per cent had a bilateral anomaly.

With a unilateral anomaly complete duplication is somewhat less frequent than incomplete. Among our cases complete duplication was revealed in 34.9 per cent and incomplete in 65.1 per cent of cases. This is in agreement with data published by Johnston (1961). Volpyan (1954) claims that complete duplication is encountered just as frequently as incomplete, while other authors think that it is rarer.

In a bilateral anomaly incomplete duplication also prevails over complete duplication. In our experience we observed bilateral ureteral splitting in 19, complete bilateral duplication in 16, and mixed in 19 cases.

The anomaly is encountered with equal frequency on the right and on the left. In our cases it was revealed in 48 per cent on the right and in 52 per cent of cases on the left. According to some authors, however, there is a clear predominance of the anomaly on the left side. For instance, among 381 cases Zschoch (1962) revealed duplicated kidneys on the right in 146, on the left in 184, and in the remaining cases the anomaly was bilateral.

It is our experience that duplicated kidneys with high ureteral splitting are more frequent. The ureter was split in the upper third in 137, in the middle third in 61, and in the lower third in 27 cases. Ermolin (1964) encountered splitting of the ureters in the upper and lower thirds most frequently.

Blind termination of the ureter in the proximal or distal segments is a rare occurrence. In the literature only occasional cases, no more than 30, are described. The same applies to the blind termination of one of the branches of a split ureter of a duplicated kidney, although Gojanna and Green (1945) found

four such anomalies among 131 duplicated kidneys, while Ferber (1963) found two among 100 cases. Among our cases of 638 duplicated kidneys we found two blind terminations of an ureter of a duplicated kidney in the distal and five in the proximal segment (a total of 1.1 per cent).

According to Blundon and Lane (1960), no less than 500 cases of ectopia of the ureteral orifice could be found in the literature by the time their work was published. In later years we found reports of another 83 cases of the anomaly in the literature, so that their total number is evidently close to 600. It should be noted that Hinman (1962) believes that duplicated kidneys account for 80 per cent of all cases. We possess information on 17 (2.8 per cent) cases of ectopia of the ureteral orifice of a duplicated kidney. All were found in females. According to Boeminhaus (1954), ectopia of the orifice is encountered five times more frequently among females than among males. This is possibly linked with the more complex diagnosis of the anomaly in males.

Ureterocele of the ureteral orifice of a duplicated kidney is also rarely encountered. Among 638 patients with duplicated kidneys we had 32 (5.0 per cent) with this abnormality. Most frequently it corresponded to an accessory ureter (accessory ureter in 22, main ureter in 4, and common ureter in 6 cases). This developmental anomaly is usually encountered in females. This is in agreement with Campbell's (1961) findings; he noted that the incidence of ureterocele was three times higher in girls than in boys. According to Lowsley and Kirvin (1956), it is bilateral in 15 per cent of cases.

The combination of these developmental anomalies of the ureter with a duplicated kidney are no longer a rarity. In our experience there were 56 such combined anomalies among 638 cases of duplicated kidneys (8.9 per cent).

Determination of the incidence of duplicated kidney with more than two ureters is of some interest. We had five patients with three pelves and three ureters and one patient with four ureters of one kidney.

Among our cases of duplicated kidneys 16 per cent had other developmental anomalies (Kalenkovich, 1970); there were seven patients with anomalies of the urinary system (lumbar dystopia of a duplicated kidney in three, its hypoplasia in one, hypoplasia of the other kidney in two, malposition of the lower segment of the ureter in one), one patient with duplication of the vagina, 32 with developmental anomalies of the bones (spina bifida in 27, lumbarization of the first sacral vertebra in one, six lumbar vertebrae in one, rotation of the spine in the lumbar segment in three), one patient with visceroptosis, and one patient with spina bifida combined with a two-horned uterus.

A duplicated kidney may often cause pain. In such cases the most thorough urological examination will fail to reveal a disease. In our experience the anomaly itself was the cause of pain in 8.3 per cent of cases. Only in 7.2 per cent of patients did the duplicated kidney produce no clinical signs and so was a chance finding during examination.

Diseases of a duplicated kidney are relatively frequent. We found them in 84.5 per cent of our cases. Kalenkovich (1970) studied the incidence of diseases of a duplicated kidney among our patients. It was established on the basis of his investigation that, as compared to normal kidneys, a duplicated kidney is affected with pyelonephritis and hydronephrosis more frequently, and with urolithiasis less frequently. Pathological mobility of a duplicated kidney is encountered no more frequently than that of a normal kidney, while tuberculosis and tumour are less frequent.

Thus, primary pyelonephritis proved to be the most common disease. It was revealed in 21.8 per cent of patients. In total, pyelonephritis was found in 64.6 per cent of patients, because it often occurred as a complication of urolithiasis, hydronephrosis or some other disease. In our clinic pyelonephritis was diagnosed in 53 per cent of patients with normally developed kidneys. Urolithiasis in duplicated kidneys was encountered in 20 per cent of cases, but it was observed in 40.9 per cent of patients with normally developed kidneys. Hydronephrosis in duplicated kidneys and associated developmental anomalies of the ureter was found in 24.1 per cent of patients, whereas its frequency in patients with normally developed kidneys was 7.5 per cent. Pathological mobility of an anomalous kidney was revealed no more often than of a normally developed kidney (14.4 and 13.7 per cent, respectively). Tuberculosis was much less frequent than in a normally developed kidney (2.6 and 8.4 per cent, respectively). Tumour proved to be the rarest disease (0.4 per cent in duplicated kidney and 3.6 per cent in normally developed kidneys).

It is interesting to note that 36.6 per cent of our cases were found to have a disease of the contralateral kidney, among which pyelonephritis was prevalent (21.3 per cent), with pathological mobility, hydronephrosis, tuberculosis and urolithiasis also being observed.

Judging from the experience of our clinic, usually the whole kidney or only its lower half is involved in a pathological process. Isolated affection of the upper part of such a kidney is a rare occurrence. Affection of the whole kidney is most characteristic of pyelonephritis, and of the whole kidney or its lower part of tuberculosis. Involvement of the lower part of the kidney prevailed in urolithiasis and hydronephrosis. Involvement of the upper

part is often attended with an anomaly of the terminal ureteral segment, such as blind ureteral termination, ectopia and ureterocele. Vesicoureteral reflux is quite often revealed in a duplicated kidney (17.8 per cent). Gimpelson (1949) noted the prevalent involvement of the lower part of the duplicated kidney. In the practice of Ratner et al. (1961) affection of the upper part of a duplicated kidney was found in 25 per cent, of the lower part in 25 per cent, and of the whole kidney in 50 per cent of cases.

SYMPTOMATOLOGY, CLINICAL COURSE AND RECOGNITION OF DUPLICATED KIDNEYS

According to the clinical course, two groups of duplicated kidneys can be distinguished: (1) asymptomatic and without any diseases, and (2) with pathological phenomena but without diseases.

An asymptomatic anomaly is rarely encountered. We observed it in 7.2 per cent of our cases. The duplicated kidney was found accidentally during urological examination for a disease of the other kidney or in making a differential diagnosis with diseases of the abdominal organs.

A duplicated kidney was often the cause of pain. We noted this in 8.3 per cent of cases. According to Volpyan (1954), such symptoms were revealed in 8.9 per cent of patients who had duplicated kidneys unaffected by disease. In addition to pain the anomaly can also cause haematuria.

Pain in a duplicated kidney varies in intensity, from mild dull pain to the severest attacks of renal colic. It is usually localized in the lumbar region, less frequently in the abdomen. It may be continuous, in which case it is of a dull character, or it can occur periodically either dull or in severe bouts. Sometimes the localization of pain is so atypical of a kidney that the clinical manifestations of the duplicated kidney are mistaken for signs of appendicitis or inflammatory diseases of the female genitals.

Quite a few of our patients with duplicated kidneys noted that at first they felt dull or sharp pain at times, but then it changed to continuous pain.

There are indications in the literature that in some cases a duplicated kidney may be the cause of profuse haematuria. We noted haematuria in one of our cases.

K., a 20-year-old man, was admitted to the clinic on July 2, 1962. He complained of dull pain in the right abdomen and small of the back occurring periodically over the previous two years. Before he had been admitted to the clinic for examination, profuse haematuria suddenly occurred in addition to the pain. The kidneys could not be palpated. Pasternatskii's sign was negative. Blood tests showed no abnormalities. Urinalysis: acid reaction,

specific gravity 1021, no protein, 5-6 leucocytes and 6-8 erythrocytes in field of vision. Excretory urography demonstrated a normal urinary tract on the left and two pelves and two ureters on the right. Pyelography revealed complete duplication of the right ureter. There was nothing to indicate a disease of the kidney or other organs. Subsequent follow-up also failed to reveal any disease of the kidney.

Pain and haematuria in cases of a duplicated kidney can apparently be linked with the disorders in urodynamics of the upper urinary tract which occur with this developmental renal anomaly. Our examination of the urinary tract of a duplicated kidney by means of urokymography showed that each ureter contracts in its own rhythm asynchronously with the other; arrhythmic contractions, lateral displacement of the ureters, changes in the diastole and prolongation of the systolodiastolic complex are often encountered. This can be explained by the asynchronous contractions of the ureters, with the jolting of one ureter against the other increasing them. In some cases counter-contractions of the ureter occur. All these changes are conducive to disturbances in the passage of urine, which may at first occur periodically and later become permanent. These dyskinetic phenomena and the urinary stasis consequent upon them may be the cause of pain and haematuria in individuals with duplicated kidneys. Later the same causes promote the development of diseases. Marked dyskinesia of the urinary tract in this anomaly should be regarded as a very early sign of the diseases which are usually associated with urinary stasis, i.e. hydronephrosis, pyelonephritis and nephrolithiasis.

Campbell's (1967) work provides proof of the complex disorders of urodynamics in cases of double kidneys and duplicated ureters. By means of cineurography he showed that the ureteral segments at the orifice contract independently of the common ureter, the ureteral contents are retained at the orifice, contractions and spasms of the ureters in complete ureteral duplication are asynchronous, and that there is a vesicoureteral reflux.

The diagnosis of duplicated kidneys is based mainly on the findings of instrumental and X-ray examination.

Cystoscopy makes the diagnosis of a duplicated kidney easy when an accessory ureteral orifice in the bladder is seen, but this is so only in cases of complete duplication of the kidney. A duplicated kidney can be suspected and looked for if alternately clear and then cloudy urine is ejected from the orifice. This is also true of chromocystoscopy when the urine discharged from the orifice is alternately clear and stained. This symptom is sometimes produced in catheterization of the ureter.

Sometimes the outlines of an elongated, spindle-shaped kidney on scout radiographs suggest this developmental kidney anomaly,

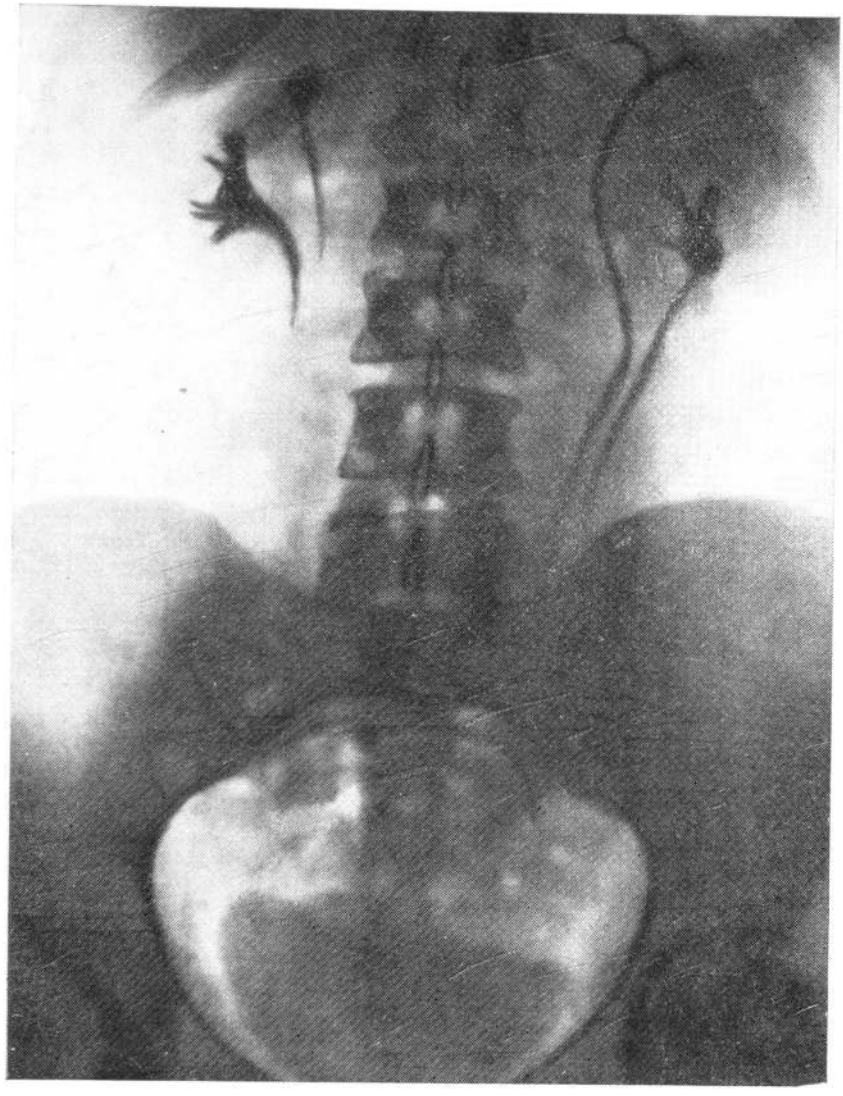


Fig. 90. Case R. Duplication of both kidneys. Excretory urography

but such an assumption should be confirmed without fail by other methods of X-ray examination.

Excretory urography is one of the main methods in the diagnosis of a duplicated kidney, making recognition of the anomaly possible in most cases. When the kidney is functioning well both pelves and both ureters are clearly visible on radiographs, and their morphological features and the function of each part of the kidney can be determined (Fig. 90).

This examination, however, does not always help in making a correct diagnosis, because on some radiographs the accessory pelvis is not clear, the detailed structure of the pelvis cannot be seen well, it is not always possible to see both ureters, and

the character of the duplication and the level of ureteral splitting very often cannot be determined. Cystoscopy is able to provide a more accurate picture. For instance, if one ureteral orifice is found in the bladder and radiographs taken during excretory urography show two pelves and two ureters, then a diagnosis of incomplete duplication of the kidney can be made. In this case, however, the level of the splitting is also not determined.

The correct diagnosis of a duplicated kidney is made more often by means of infusion urography. But this method also does not always show the details of the pyelo-calyceal system clearly or reveal the character of the duplication of the kidney and the level of ureteral splitting. In addition, the findings of excretory urography are sometimes insufficient for making a differential diagnosis between a duplicated kidney and diseases of an anomalous kidney.

Retrograde pyelography yields more convincing proof in favour of the renal anomaly and helps to recognize or rule out diseases. This method is effective in cases of poor function of a part or the whole of the duplicated kidney, when excretory urography cannot help in the diagnosis. Retrograde pyelography also shows an interureteral reflux during which the contrast medium flows from one ureter into the other non-catheterized ureter. Because of the reflux the structure of both pelves and ureters can be seen on the radiographs (Fig. 91).

Retrograde pyelography makes it possible to determine the type of duplicated kidney, the level of splitting, and the morphological features in almost all cases. The only exceptions are those in which the catheters cannot be advanced into the ureters or when the examination techniques are faulty.

A typical pyelogram of a duplicated kidney shows the upper pelvis as a 'carnation' with two, or less frequently with one or three, calyces (Fig. 92). Below it is the second pelvis, usually larger, and sometimes still another pelvis (Fig. 93). The lower and middle calyces are normal, while the upper calyx of this pelvis is situated transversely as if it is flattened. Sometimes it is rudimentary or absent. In bilateral duplication the duplicated pelves on each side are usually of different shape.

It is sometimes difficult to make a diagnosis of a duplicated kidney when the contrast medium fills only one pelvis and its ureter. In such cases 'indirect X-ray signs' of a duplicated kidney are very helpful. For instance, if there is an image of the upper pelvis and its ureter, the diagnosis may be made from its characteristic shape, its high position in the lumbar region and, if the kidney is outlined, according to the disproportionate position of the pelvis in relation to the entire mass of the kidney. When the lower pelvis and its ureter are seen on the radiograph,

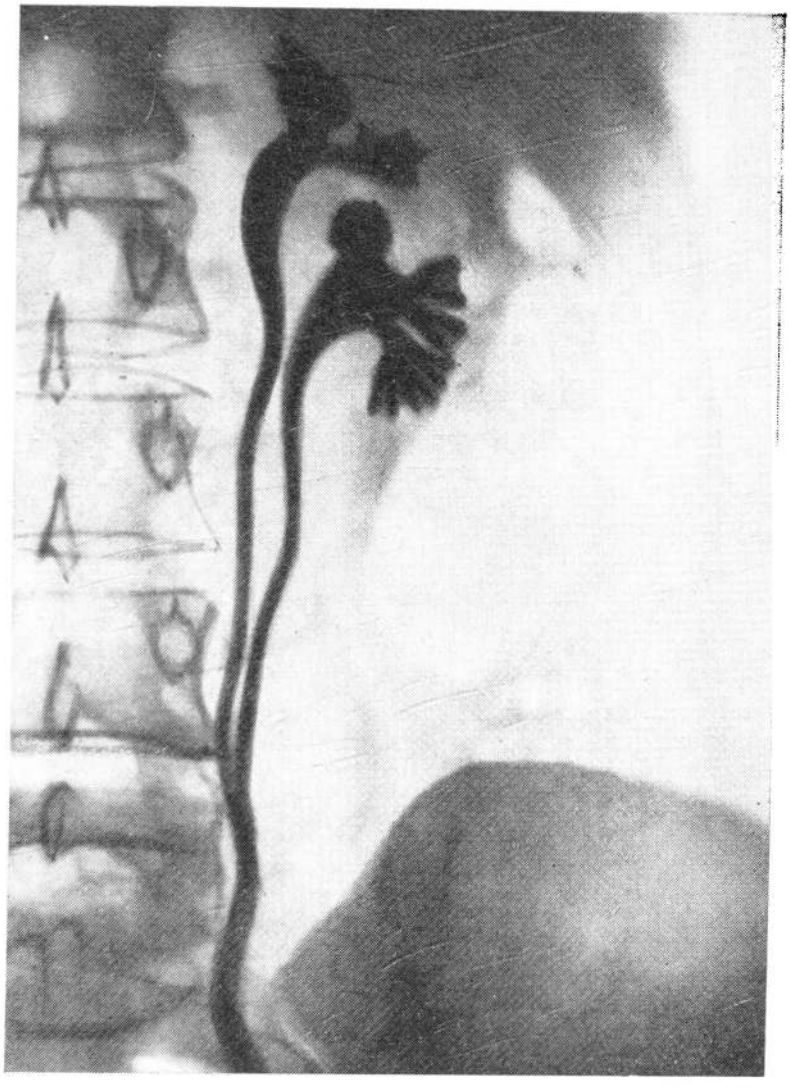


Fig. 91. Case K. Incomplete duplication of the left kidney. The catheter is introduced into the upper renal pelvis, but an image of the whole urinary tract is produced. Retrograde pyelography

the diagnosis of a duplicated kidney can be made on the basis of the defective upper calyx and the non-conformity of the urinary tracts and their low position in relation to the outlines of the kidney. In such cases diagnostic pneumoretroperitoneum is undertaken to obtain a clearer image of the kidney.

Pneumoretroperitoneum is a valuable method in the diagnosis of duplicated kidney on the basis of indirect signs. In such cases an elongated or spindle-shaped kidney is seen on which grooves corresponding to the separation of the kidney into two parts are seen. This method of examination, however, helps only in

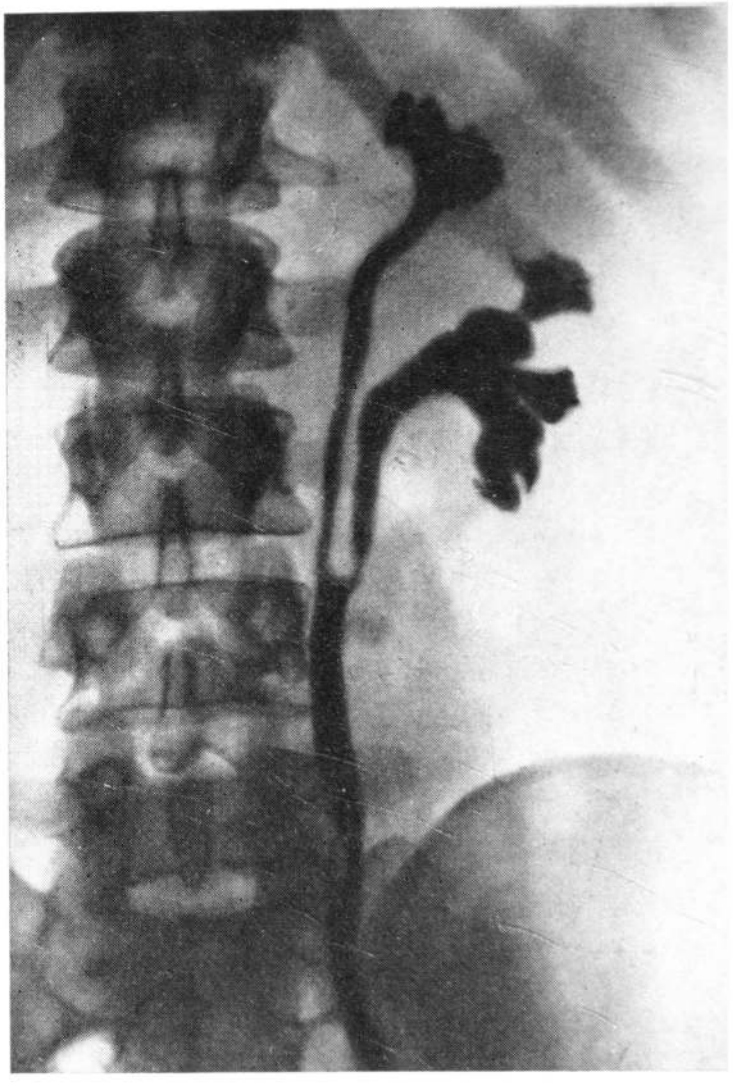


Fig. 92. Case K. Duplication of the kidney with ureteral splitting. Retrograde pyelography

presuming the diagnosis of the anomaly, because it does not show the duplicated upper urinary tract. But it does give a more exact idea of the shape and position of the organ, which may be valuable in differential diagnosis with diseases of the kidney and other organs and before operation. The structure of a duplicated kidney and its urinary tracts is shown particularly clearly on radiographs taken when pneumoretroperitoneum is combined with excretory urography or retrograde pyelography (Fig. 94).

In some cases, the findings of excretory urography and tomography, showing the outlines of a duplicated kidney and duplicated urinary tracts, are sufficient for confirming the diagnosis

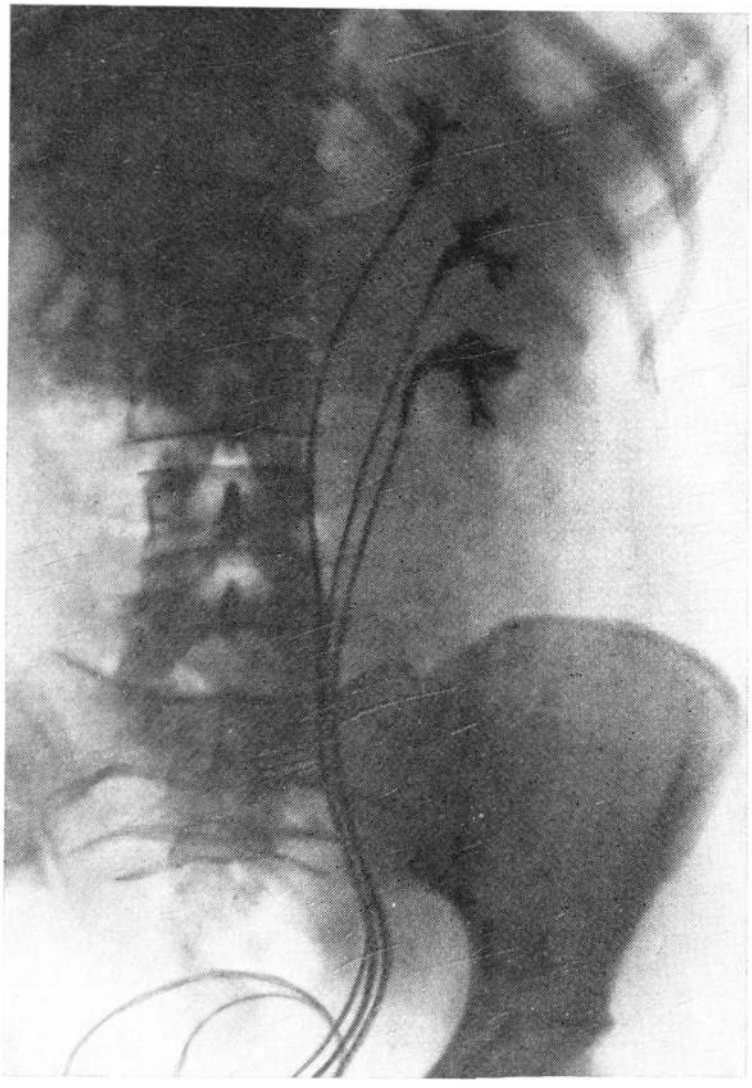


Fig. 93. Case K. Complete triplication of the upper urinary tract. Retrograde pyelography

of the anomaly. We found this examination to be particularly valuable in ectopia of one of the ureters, when it was important to gain information on the structure of the kidney prior to operation. The levels of tomographic sections in these cases are the same as those used in the examination of normal kidneys.

Indications for the use of renal angiography in cases of duplicated kidneys are very restricted (Lopatkin, 1967). This method allows for the study of the angioarchitectonics of each part of the kidney and gives an idea of their functional and morphological condition. It is used in cases difficult for diagnosis, when the methods discussed above yield no data.

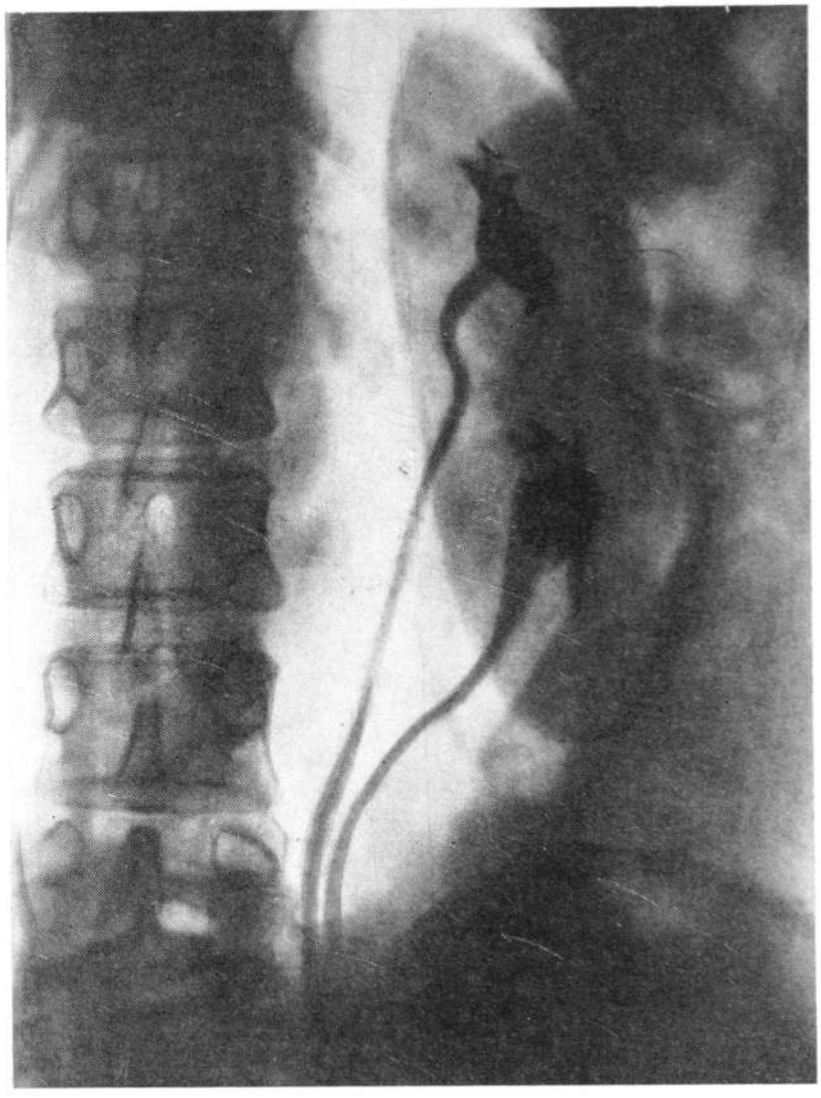


Fig. 94. Case K. Duplication of the left kidney. Pneumoretroperitoneum and excretory urography

Occasional reports may be found in the literature of the use of urokymography for studying the dynamics of the upper urinary tract of a duplicated kidney (Gregoir, 1953; Gorshkova, 1966).

In our use of this method we found that the contractions of the upper urinary tracts of a duplicated kidney were usually arrhythmic but physiologically directed, sometimes retrodirected. Contractions of the lower pelvis were weaker than those of the upper pelvis. The alteration of waves with the usual amplitude of contractions was somewhat slower. In all cases, both ureters contracted with a different rhythm and presystolic contractions

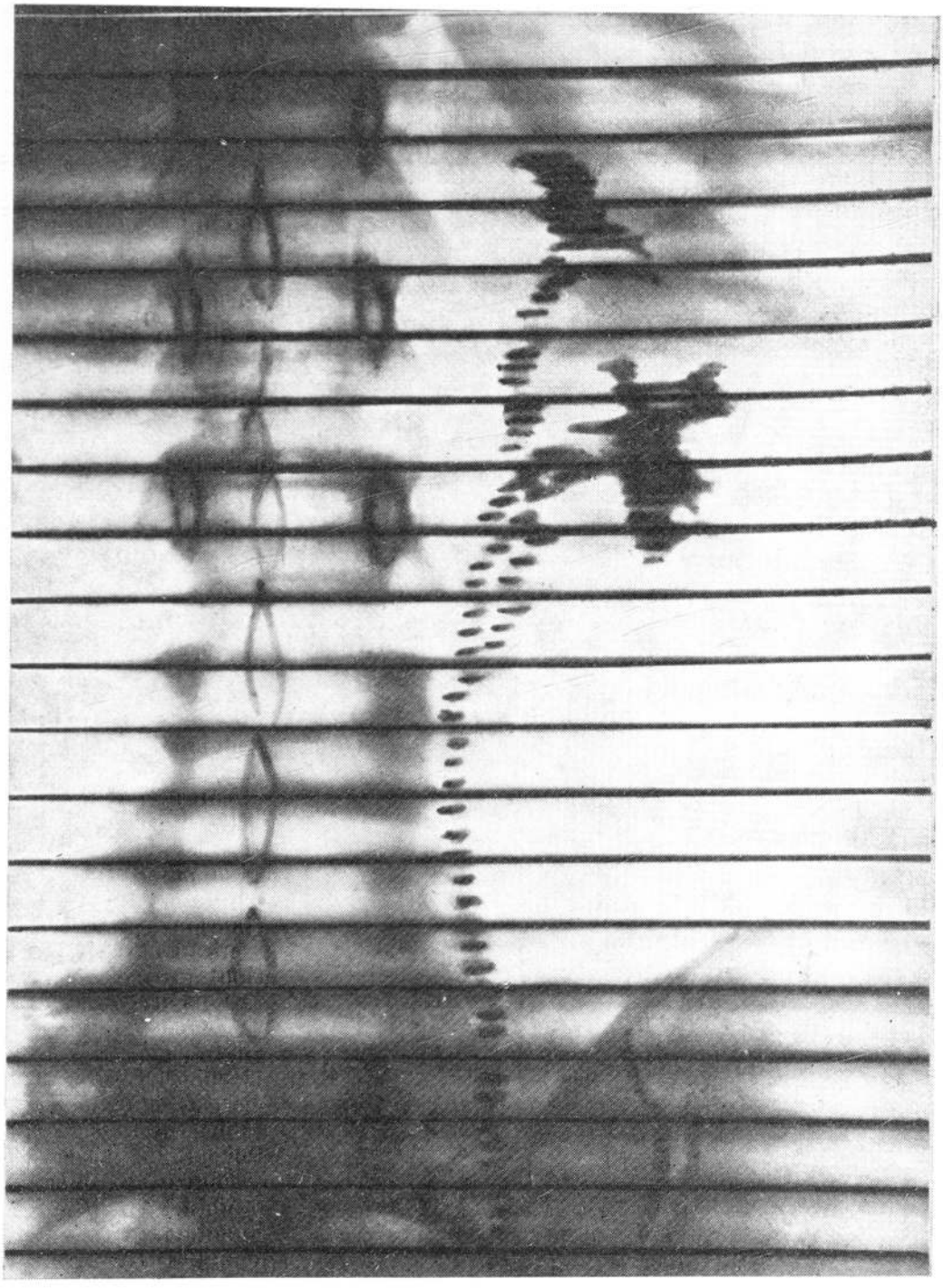


Fig. 95. Case S. Duplication of the left kidney. Urokymogram

were observed where they came in contact. Lateral displacement of the ureters and deformities of their image at the site of their contact with the pelvis were often seen (Fig. 95). These changes

are evidence of dyskinetic phenomena which are often observed in the upper urinary tracts of a duplicated kidney and are evidently the cause of pathological disturbances and signify the onset of disease.

Radioisotope methods of examination do not always help in the diagnosis of a duplicated kidney. They only help to define precisely the general outlines of the kidneys and the function of the whole kidney.

A duplicated kidney has to be differentiated from other developmental renal anomalies (hypoplasia, fused kidneys and dystopia of the organ) and from diseases of the kidneys and other organs. The differential diagnosis in such cases is often quite difficult and errors cannot be avoided.

If only the image of the upper rudimentary pelvis of a duplicated kidney is seen on the radiographs, it is sometimes mistaken for a hypoplastic kidney. The findings of pneumoretroperitoneum, which help to reveal the discrepancy in the positions of the kidney mass and the urinary tract, are of great value in the diagnosis. Tomography in combination with excretory urography and pneumoretroperitoneum prevent errors in exactly determining the side of the duplication with ectopia of the ureteral orifice, and in revealing a combination of several anomalies in one kidney (duplication of a dystopic kidney, duplication of a horseshoe kidney, etc.). The use of renal angiography in cases of difficult diagnosis is justified.

In some cases it is very difficult to differentiate a duplicated kidney from diseases of other organs. In one of our cases of a tumour in the retroperitoneal space, a duplicated kidney created the picture of a horseshoe kidney: dull lumbar pain, a tumour palpated in the region of the umbilicus, duplication of the urinary tract on the right, and dystopia of the kidney on the left. The principal disease proved to be retroperitoneal lymphosarcoma.

In patients with a duplicated kidney the symptoms of a tumour in the intestine or female genitals are often attributed to a renal anomaly as a result of which improper treatment is prescribed.

In other cases abnormalities, in fact resulting from a duplicated kidney, call for unjustified operations only during which is a correct diagnosis belatedly made. Thorough preoperative examination, however, makes it possible to avoid errors.

L., a 33-year-old man, was admitted to the clinic on February 3, 1964 with the diagnosis of tumour of the kidney. For the previous year he had experienced attacks of pain in the right lumbar region. A nodular and tender lower pole of the right kidney was palpated. Blood test: haemoglobin 83 per cent; leucocytes 7700; ESR 39 mm/hour. Urinalysis showed normal values. Duplication of the right kidney with high splitting of the ureter was revealed by pyelography. The lower pelvis was rotated slightly down, its lower calyx could not be seen clearly. Diagnostic pneumoretroperitoneum showed an

enlarged right kidney with irregular contours of the lower pole. Tumour of a duplicated kidney was suspected. It was found during surgery that a thick infiltrate continuous with a tumour of the caecum was fused to the lower pole of the kidney. Hemicolectomy with ileotransversostomy was carried out. Histological examination of the tumour showed it to be adenocarcinoma of the caecum.

Thus, when a diagnosis of duplicated kidney is made, it is necessary to rule out the existence of some disease of organs of the abdominal cavity or a disease of the duplicated kidney, the manifestations of which may be mistaken for the symptoms of the renal anomaly.

PECULIARITIES OF THE SYMPTOMATOLOGY, COURSE AND RECOGNITION OF CONGENITAL DEFECTS OF THE URETER OF A DUPLICATED KIDNEY

Blind termination of the ureter of a duplicated kidney in the distal segment may produce no symptoms and be revealed by chance during examination or as a result of injury. In a case reported by Kremnev (1938), this anomaly was diagnosed in a patient with subcutaneous rupture of a large ureterohydronephrosis. In most cases, however, the anomaly apparently produces symptoms of secondary diseases (hydronephrosis, pyelonephritis, nephrolithiasis) associated with impaired passage of urine. These patients note occasional swelling in the abdomen, in the inguinal region, or in the region of the bladder or vagina corresponding to the site of the blind ureteral termination. Urinalysis sometimes reveals haematuria or pyuria. These clinical manifestations are noted over a period of many years. The following case is given in illustration.

D., a 65-year-old man, was admitted to the surgical department on October 1, 1956, complaining of dull pain in the lower abdomen and a tumour in the left inguinal region. He had been ill for about 30 years. He experienced unpleasant sensations now and again, as well as dull pain in the left abdomen and small of the back. Three years earlier he had found a swelling in the left inguinal region which was taken for an inguinal hernia by doctors. In the previous three weeks dull pain in the lower abdomen and painful micturition had developed. A tender sausage-shaped infiltrate was palpated in the left inguinal region along the length of the inguinal canal. A tender thickening was palpated through the external opening of the inguinal canal. Incarcerated inguinal hernia on the left side was diagnosed, but during surgery the inguinal canal was found to be free. There was an infiltrate above the iliopsoas muscle containing a cavity with smooth walls and clear fluid.

The patient was sent to the urological department. Urinalysis revealed haematuria, pyuria and albuminuria. Cystoscopy could not be performed because of the small volume of the urinary bladder. The kidneys were not outlined on scout radiographs and no shadows of concrements were seen. During excretory urography, a dilated left pelvis and ureter were demonstrated on the radiographs. Cystograms showed a very small urinary bladder

with irregular outlines. Methylene blue solution infused into the bladder was not discharged from the wound; the patient's condition deteriorated progressively and 24 days after admission to the clinic he died.

Pathological diagnosis: carcinoma of the urinary bladder with predominant growth on the right; compression by the tumour of two ureteral orifices in the bladder, more on the right; dilatation of both ureters; bilateral hydronephrosis with pyelonephritis. The left kidney is duplicated and has one accessory ureter which is also dilated the same as the corresponding pelvis. The accessory ureter terminates blindly in the inguinal region and communicates with the operative wound.

This patient had had symptoms of the anomaly for a lengthy period of time, up to thirty years. A peculiar cyst of the accessory ureter was palpated and mistaken for an inguinal hernia. The development of a concomitant severe disease, a tumour of the urinary bladder compressing the ureteral orifices in the bladder, caused progression of the disease and death. The renal anomaly was not recognized before operation. It was found that the accessory ureter of the duplicated kidney terminated blindly in the tissue of the inguinal region by the urinary bladder.

This developmental anomaly is quite difficult to diagnose and is usually revealed during operation or at autopsy. But if it is borne in mind, its diagnosis can be made prior to operation, as was the case in one of our patients.

B., an 11-year-old girl, was admitted to the clinic on September 7, 1966, complaining of pain in the left lumbar region, frequent and painful micturition and subfebrile temperature. She had been ill for five years. Treatment for pyelonephritis had no marked success. The region of the left kidney was tender to palpation. A blood test showed no abnormalities. Urinalysis revealed 10-15 leucocytes and occasional erythrocytes. Urine culture produced growth of coli bacillus. Urography showed good functioning of both kidneys and normal pelves. Chromocystoscopy revealed normal ureteral orifices in the bladder, one on each side. A swelling 2.5 cm in diameter was seen below the left orifice. Indigo carmine was discharged from both ureters in a full stream in the fifth minute of observation. Vesicoureteral reflux was noted on the left side of the cystogram, a filling defect on the left was seen in a small filling of the bladder. Duplication of the left kidney with blind ending of the ureter at the bladder was suspected. Endovesical electrocoagulation of the structure protruding into the bladder was conducted; purulent fluid was discharged from the opening thus formed.

The following diagnosis was made: total duplication of the left kidney with ureter terminating blindly under the mucosa of the urinary bladder. Vesicoureteral reflux into the main ureter of the duplicated kidney. Pyelonephritis. Surgery was performed: left ureterectomy with heminephrectomy of the upper part of the duplicated kidney, Gregoir's anti-reflux operation on the lower ureter. The upper part of the kidney was contracted, it had a small pelvis and an ureter obliterated in the prepelvic segment. The girl recovered.

In this case recognition of the swelling in the urinary bladder and its electrocoagulation helped to reveal the blind termination of the ureter of the duplicated kidney. It was found that the

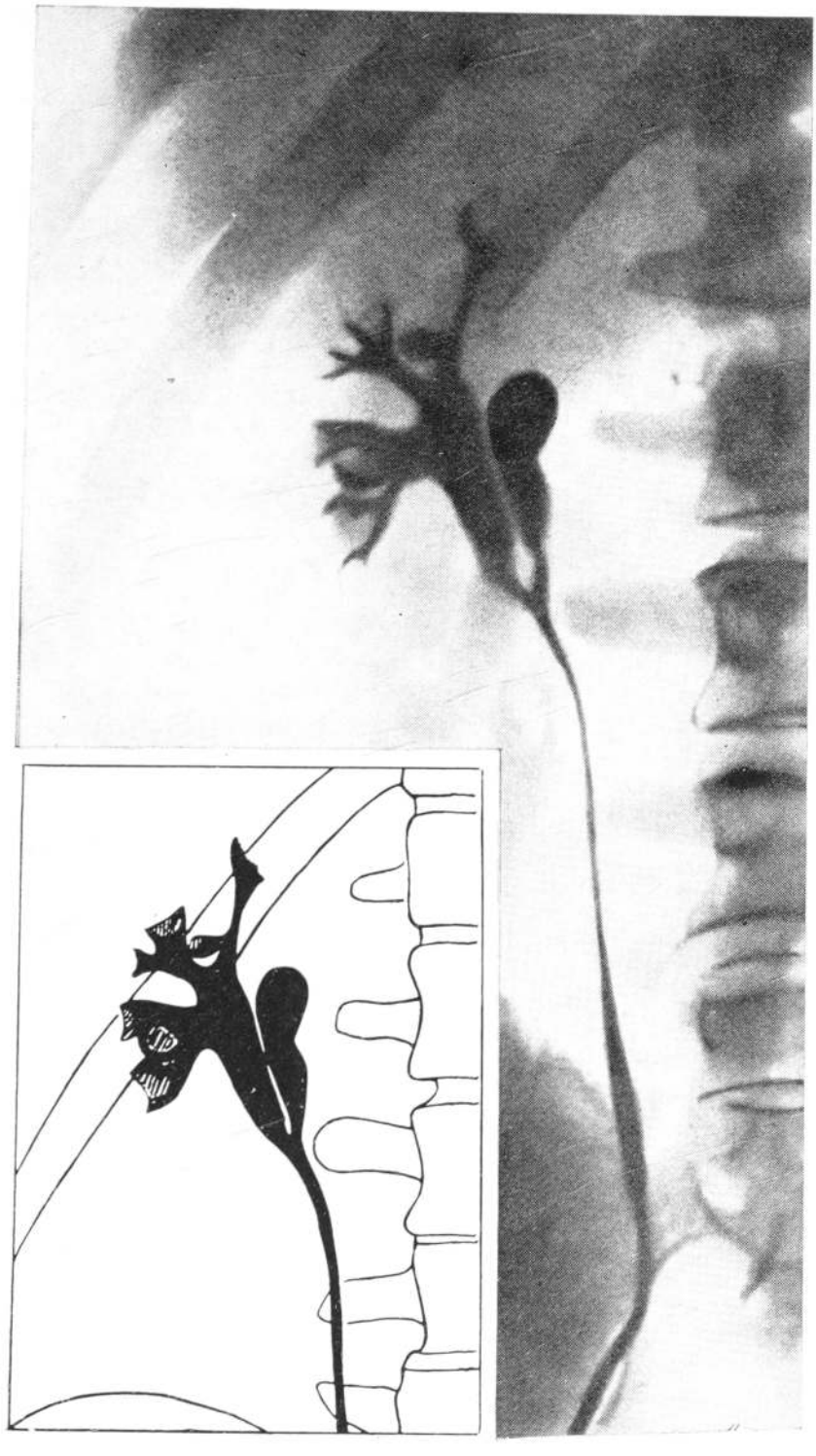


Fig. 96. Case A. Duplication of the right kidney with splitting of the ureter. Blind termination of the accessory ureter at the kidney

accessory ureter of the duplicated kidney terminated blindly in the vesical submucosa and was obliterated proximally at the pelvis.

In cases of poor evacuation of the contents, blind termination of the proximal segment of the ureter of a duplicated kidney produces an indefinite clinical picture: periodical pain in the abdomen or small of the back, frequent or painful micturition, pyuria, microscopical haematuria and proteinuria. Pain is dull or, less frequently, of the type of renal colic. Sometimes this anomaly causes a clinical picture which is mistaken for the symptoms of appendicitis or adnexitis. A most typical clinical picture in such cases is that of chronic inflammatory disease of the urinary tract.

Diseases in developmental anomalies of the ureter usually follow a protracted chronic course. The discovery of a second ureteral orifice during cystoscopy and a blindly terminating ureter during retrograde pyelography are of major importance in making a diagnosis (Figs. 96 and 97).

Retrograde pyelography also helps in the correct diagnosis of a blindly terminating ureter when there is only one ureteral orifice in the bladder and the ureter is split into two branches, one of which has a blind ending. In such cases, because of the interureteral reflux, the contrast medium introduced into one ureter produces a picture of the whole duplicated kidney and the blindly terminating ureter on the radiographs. The following case serves as an illustration.

E., a 37-year-old man, was admitted to the clinic on February 16, 1971, complaining of dull pain in the left lumbar region. He had been ill for seven years. Dull pain appeared from time to time in the left lumbar region, body temperature was sometimes subfebrile and micturition was frequent and painful. Antibacterial treatment brought temporary relief. The lower pole of the kidney was felt in the left hypochondrium, and palpation of this region was painful. Pasternatskii's sign was weakly positive on the left. Blood test showed normal values. Urinalysis: acid reaction; specific gravity 1017; protein 0.33 pro mille; leucocytes 35-40 in field of vision. It was seen on chromocystoscopy that the urinary bladder had two orifices, one on each side, from which indigo carmine was discharged in the third minute. Excretory urography showed good functioning of both kidneys, and the pelves and ureters were normal. Retrograde pyelography showed a duplicated left kidney with spitting of the ureter in the lower third; the accessory ureter terminated blindly at the prepelvic segment of the main ureter (Fig. 98).

During retrograde pyelography the catheter was advanced along one of the ureters to a height of 20 cm, but because of interureteral reflux the contrast medium also filled the second ureter. The following diagnosis was made: duplication of the left kidney with splitting of the ureter in the lower third, accessory ureteral branch terminating blindly at the pelvis, ureteritis. After adequate preparation surgery for ureterectomy of the blind branch of the accessory ureter was performed. The postoperative period was uneventful. The patient recovered.

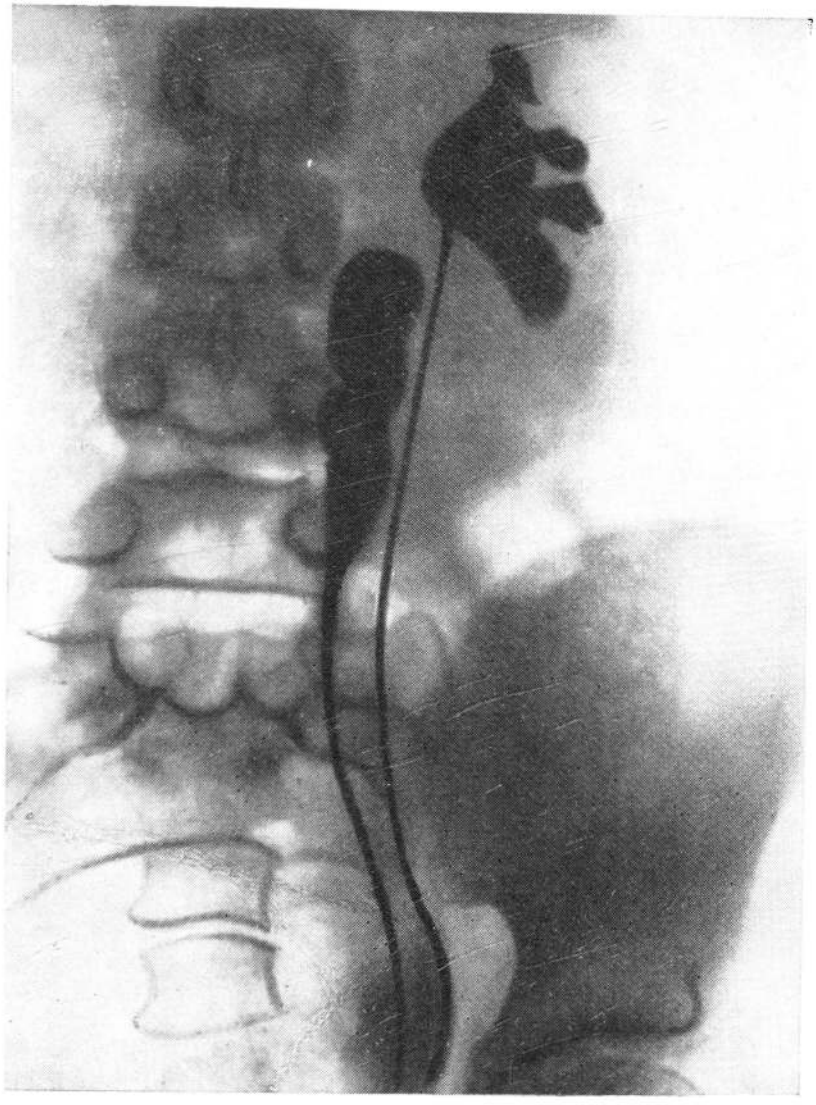


Fig. 97. Case Z. Complete duplication of the left kidney. Blind termination of the accessory ureter at the kidney

Ectopia of the orifice of the ureter of a duplicated kidney is usually revealed in children because of the peculiar clinical picture, but it can be observed much later. In our experience, for example, this developmental malformation was found in 17 patients between the ages of 2 and 53 years.

The clinical picture of ureteral ectopia is determined to a large extent by the site of the ureteral orifice, the symptoms produced by the organs into which the urine escapes, the condition of the kidney, and infection. According to the position of the ureteral orifice, it is expedient to distinguish two groups of patients with ectopia of the ureter, as proposed by Ermolin (1964). The

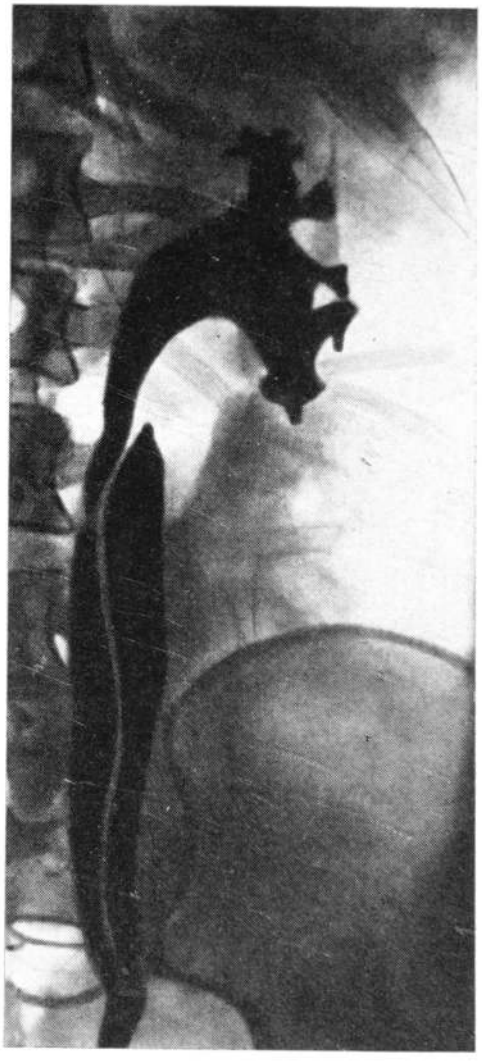


Fig. 98. Case E. Duplication of the left kidney with splitting of ureter and blind termination of the accessory branch at the kidney. Retrograde pyelography

first group consists of patients with suprasphincteral ectopia, when the ureter drains into the bladder. The second group is composed of patients with extrasphincteral ectopia when the ureter drains into the posterior urethra up to the external sphincter. In the second case the urine flows into the entrance to the vagina, into the uterus, anterior urethra, etc. This classification is simple and convenient in clinical practice.

Suprasphincteral ectopia of the ureteral orifice is encountered less frequently than extrasphincteral ectopia. We found it in only two of our cases, whereas extrasphincteral ectopia was discovered in 15 cases. In suprasphincteral ectopia the urine drains into the urinary bladder and is retained there by the sphincter, and because of this incontinence of urine does not occur. These patients may have no complaints and the anomaly is revealed by chance.

The clinical picture of suprasphincteral ectopia of the ureteral orifice may be composed of the following symptoms: periodical dull pain in the small of the back, disturbed micturition, pyuria, microhaematuria, proteinuria, bacteriuria and positive Pasternatskii's sign. These symptoms usually occur because of the poor passage of urine and infection which develops with time when the anomalous orifice is narrow; at first they occur occasionally, later to become permanent. This is why the disease follows a chronic course and after some time is complicated by pyelonephritis, hydronephrosis and nephrolithiasis.

The diagnosis of suprasphincteral ectopia of the ureteral orifice is easy if the ectopic orifice is revealed during cystoscopy. Then the diagnosis of duplicated kidneys and ureters can be made from the findings of excretory urography or retrograde pyelography. If the orifices cannot be found in the usual place then the diagnosis of duplicated kidney is made first on the basis of the X-ray methods just mentioned. It becomes necessary to determine whether or not the ureter is split. This, however, is not always possible. At times the diagnosis can only be made during surgery. In other cases retrograde pyelography with the catheter advanced to a low level is applied. The presence of a poorly functioning duplicated kidney is determined by the discrepancy between the position of the pelvis and the renal mass found during pneumoretroperitoneum and retrograde pyelography.

Extrasphincteral ectopia of the ureteral orifice in cases of a duplicated kidney is marked by continuous involuntary discharge of urine but with preserved act of voluntary micturition. This is described in the literature as Pozner's symptom. It is usually encountered constantly, but sometimes it occurs only when the patient takes a vertical position, which may be explained by the accumulation of urine in the dilated ureter when the patient is in a horizontal position. When secondary diseases develop in the anomalous kidney and there is concomitant infection, these patients experience lumbar pain, which is either dull or occurs in attacks. The continuous flow of urine causes characteristic maceration of the thighs.

In extrasphincteral ectopia of the ureteral orifice Pozner's symptom is usually observed from birth. In one of our patients, however, it became apparent two years before she was admitted to our clinic. In this case the ureter evidently terminated in a cyst formed of remnants of Gartner's duct and urine was discharged only after rupture of the cyst.

As a result of the poor drainage of urine from the ectopic orifice, which is of uncommon localization and has a long and tortuous terminal ureteral segment, this developmental anomaly may be attended by secondary diseases such as hydro- and pyouretero-

nephrosis, atrophy of the kidney or nephrolithiasis. The kidney is often affected with pyelonephritis. Among 15 patients of this group we had only two in which the kidney functioned well and there were no symptoms of disease. All the other patients had symptoms of pyelonephritis and hydronephrosis. The diseases mentioned here were encountered in half of all cases of this anomaly found in the literature by Salturin (1940).

Pozner's symptom makes it possible to presume ectopia of the ureteral orifice outside the sphincter vesicae, but the diagnosis must be confirmed by means of other examination methods, because a similar clinical picture is produced by urinary incontinence in cases of a weak sphincter vesicae, ureterovaginal or vesicovaginal fistulas and neurogenous urinary bladder.

To confirm the diagnosis of this malformation, it is most important to find the ectopic orifice. With this in mind the anterior urethra, the entrance to the vagina and, if possible, the vagina, are carefully inspected. Areas difficult for access are examined with an urethroscope, cystoscope and colposcope. Ectopia of the ureteral orifice can be suspected if one of the orifices cannot be found in the bladder. According to Epshtein and Vainshtein (1936), in ectopia of the ureteral orifice into the seminal vesicle the corresponding part of the cystic wall is sometimes distorted. One rarely has a chance of seeing the ectopic orifice, however. We had this chance only in eight of 15 cases.

To differentiate ectopia of the ureteral orifice into the anterior urethra it is expedient to use the simple test of indigo carmine administered into the urinary bladder through a catheter. A gauze tampon is placed to the external urethral orifice and vagina. The patient is asked not to urinate and to walk for 15 to 30 minutes. If the tampon is stained blue it can be presumed that the patient is suffering from urinary incontinence as a result of a weak sphincter vesicae or a vesicovaginal fistula. If the tampon under these conditions is saturated with a clear fluid, ectopia of the ureteral orifice or a ureterovaginal fistula can be suspected.

In cases of an adequately functioning kidney, chromocystoscopy will be of some help in making a diagnosis. During this examination staining of the discharged fluid may be expected and the site of the discharge is more easily found. The part of the kidney corresponding to the ectopic orifice, however, is often altered by disease. That is why clear staining of the urine in such cases cannot be obtained and the method rarely helps in the recognition of this renal anomaly.

Excretory urography helps in diagnosing this malformation, if the kidney functions adequately. In this case the duplicated kidney is seen on the radiographs. If only one of the two orifices

is seen in the bladder, ectopia of the ureteral orifice or splitting of the ureter should be considered. Retrograde pyelography helps to differentiate them in most cases. Clearer images can be produced by compressing the ureters or by means of infusion urography. In cases of diminished function of the renal segment with the ectopic ureteral orifice, however, this examination is ineffective. It fails to reveal to which kidney the ureter with the ectopic orifice belongs.

Even when an ectopic orifice of an ureter is found, it is not always easy to determine to which kidney this ureter belongs. For this purpose some authors recommend obturating the orifice with a thick catheter or suturing it for 24 hours (Lioznov, 1957; Akseldorf, 1964). In such cases renal colic develops by which the side of the anomaly is judged, but in some such cases the renal colic is attended by an attack of severe pyelonephritis, owing to which surgery cannot be restricted to an organ-preserving

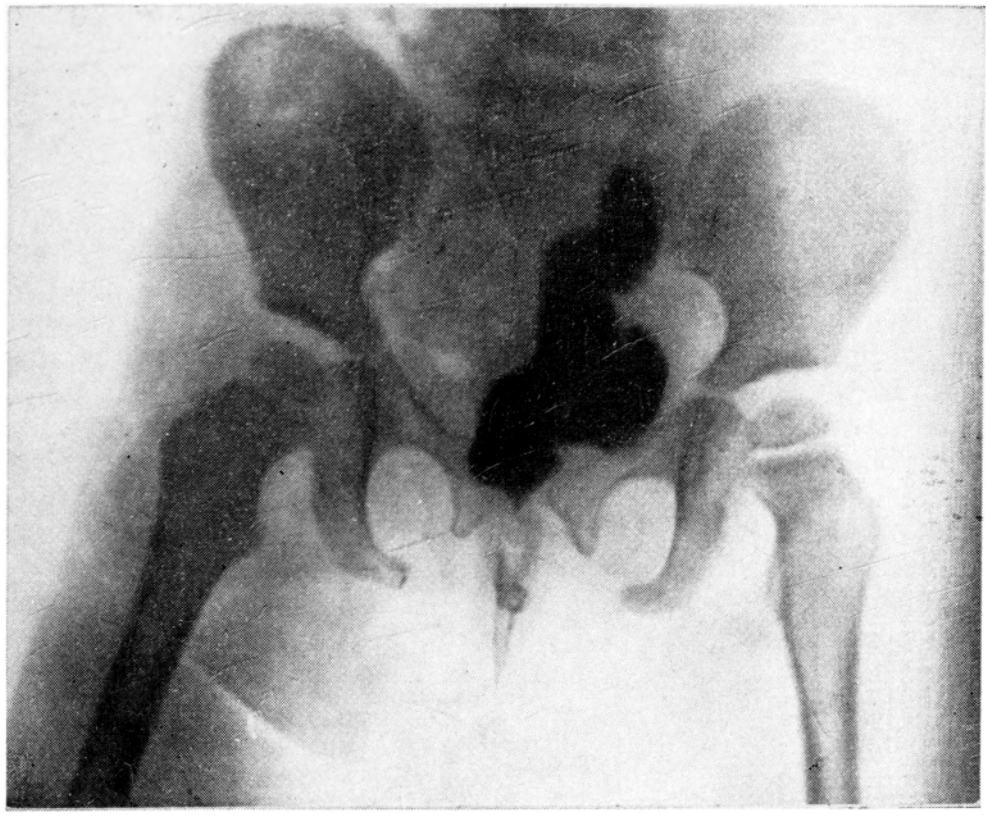


Fig. 99. Case Kh. A catheter is introduced through the ectopic ureteral orifice and retrograde pyelography performed. Duplication of the left kidney, ectopia of the orifice of the accessory ureter into the entrance of the vagina; hydronephrotic transformation of the upper part of the duplicated kidney

operation later. For this reason we do not recommend this manipulation.

It is fairly easy to make a diagnosis if the catheter can be introduced into the ectopic orifice and retrograde pyelography performed. The side of the anomaly can thus be determined exactly. Retrograde pyelography by way of this orifice helps to determine the pelvis to which the ureter with the ectopic orifice belongs. The following case serves as an illustration.

K., a 2-year-old girl, was admitted to the clinic on May 15, 1971 with continuous discharge of urine but with voluntary micturition maintained. She had had this symptom from birth. On inspection maceration of the thighs resulting from the discharged urine was noted. A pinpoint opening from which clear fluid was discharging was found in the entrance to the vagina. The fluid remained clear even after indigo carmine solution had been administered into the urinary bladder. Blood and urine tests showed no abnormalities. Excretory urography showed that the function of both kidneys was preserved; a pelvis was visible in each hypochondrium. During retrograde pyelography a normal pelvis and ureter were seen on the left. Retrograde pyelography of the ectopic ureter produced an image of a greatly dilated ureter stretching to the left kidney (Fig. 99).

The following diagnosis was made: duplication of the left kidney with ectopia of the orifice of an accessory ureter into the entrance of the vagina, ureterohydronephrosis of the upper part of the kidney and accessory ureter. Operation for heminephroureterectomy was conducted. The postoperative period was uneventful. The child recovered.

Retrograde pyelography helped to make the exact diagnosis of the anomaly in this case and determine the side on which it had developed. The side of the anomaly may also be determined by means of pneumoretroperitoneum and retrograde pyelography from the disparity in the shadow of the kidney and the shadow of the upper urinary tract. Williams and DeBaker (1962) used the findings of renal angiography for the same purpose.

It is rather more difficult to make a diagnosis and determine the side of the ectopic ureteral orifice in cases of bilateral duplication of the kidney. But in these cases as well all diagnostic doubts can be removed if the ectopic orifice is found and retrograde pyelography performed.

In one of our cases we were able to verify the diagnosis of a duplicated kidney by means of excretory urography and tomography.

When an extrasphincteral ureteral orifice is not found and also in cases of bilateral mixed duplications and bilateral ectopia of the ureteral orifices, it is difficult to diagnose this anomaly. In one of our patients with bilateral ectopia of the ureteral orifices the malformation was at first found only on one side, but inefficacy of the ureteropelvic anastomosis called for additional examination which revealed ectopia of the ureteral orifice on the other side as well. A similar case was reported by Zhukov (1957).

Sometimes, because of the latent course of this developmental malformation, it is found accidentally during surgery. In one patient treated by Baines (1940) a duplicated kidney with ectopia of the orifice of the accessory ureter into the posterior urethra was discovered during surgery for a tumour of the kidney.

The diagnosis of this anomaly should proceed as follows: detection of Pozner's symptom suggesting the anomaly; attempts to prove by means of chromocystoscopy that the discharged fluid is actually urine; determination of the site where the fluid is discharged; ruling out any association between the seeping fluid and the urinary bladder by means of the indigo carmine test; establishing by X-ray examination to which kidney the ureter with the ectopic orifice belongs and to which pelvis; exact determination of the condition of the anomalous kidney by means of the same methods of examination.

Ureterocele of the ureter of a duplicated kidney has the same symptoms as that of the ureter of a normal kidney. The clinical picture consists of the following symptoms: pain and tenderness in the lumbar region, positive Pasternatskii's sign during exacerbation of infection accompanied in these cases by a rise in body temperature and micturition disorders, pyuria, bacteriuria and proteinuria. This developmental malformation impairs the flow of urine from the ureter and therefore is very often conducive to the development of hydronephrosis, pyelonephritis and nephrolithiasis, with the corresponding clinical picture thus appearing. The disease usually follows a chronic course.

It is not difficult to recognize this developmental anomaly. The diagnosis is based on the findings of cystoscopy during which a protrusion in the wall of the urinary bladder, corresponding to the ureteral orifice, can be seen. In cases of good functioning of the kidney, the anomaly and the disease can be revealed by cystoscopy and excretory urography. If the function of the kidney is diminished, the diagnosis has to be based on indirect signs, as occurred in the following case.

Ch., a 29-year-old woman, was admitted to the clinic on May 16, 1957 complaining of dull pain in the right hypochondrium experienced over a period of 12 months. An elastic and tender lower half of the right kidney was palpated. Pasternatskii's sign was positive on the right. Blood tests showed normal values. Urinalysis: acid reaction; specific gravity 1018; protein 0.66 pro mille; leucocytes and erythrocytes covering the entire field of vision. Urine culture produced growth of enterococcus and Gram-positive diplococci. Cystoscopy revealed ureterocele on the right. Attempts to introduce a catheter into the right ureter failed. Urography showed a clear image of a normal pelvis on the left; slow radiographs showed on the right side a dilated pelvis with dilated calyces, resembling the pelvis of a duplicated kidney and located in the lower part of the shadow of the kidney.

The following diagnosis was made: hydronephrosis of the right duplicated kidney, ureterocele, pyelonephritis. Nephroureterectomy was performed.

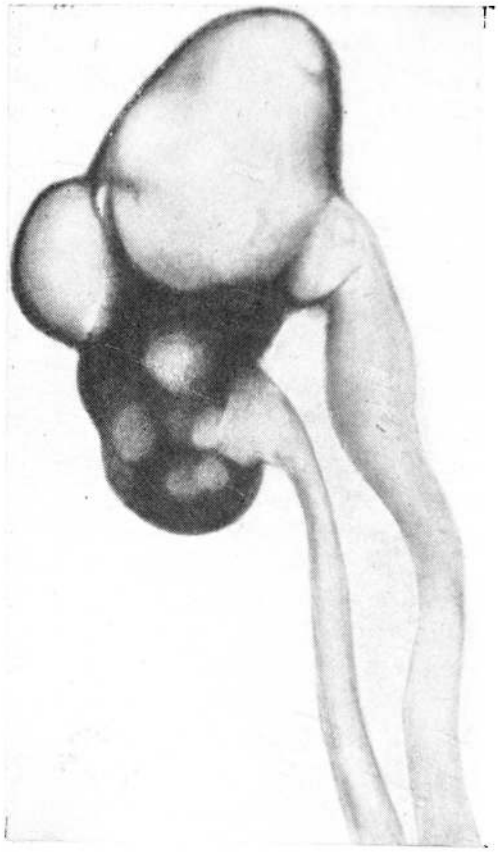


Fig. 100. Case Ch. Hydronephrotic transformation. Preparation of the removed right duplicated kidney

A duplicated kidney with splitting of the ureter in the lower third and hydroureteronephrosis with a somewhat lesser involvement of the lower part of the kidney were found during operation (Fig. 100). The patient recovered.

In order to possess an exact diagnosis prior to surgery, it would have been necessary to make an incision in the ureterocele and perform retrograde pyelography, but in this case there had been sufficient indirect signs pointing to the anomaly and the disease and indicating the need for surgery.

PECULIARITIES OF THE SYMPTOMATOLOGY, CLINICAL COURSE AND RECOGNITION OF DISEASES OF DUPLICATED KIDNEYS

In diseases of duplicated kidneys certain peculiarities are sometimes encountered which are discussed below.

Pyelonephritis. The development of this disease in people with duplicated kidneys is promoted by the frequently encountered developmental anomalies of the ureter and vesicoureteral refluxes, which cause disorders in the passage of urine from the upper

urinary tract. Judging from our cases, the whole kidney is usually involved in this disease. It is less frequent for the lower half alone to be involved and even rarer for the upper part. However, the extent of changes in involvement of the whole duplicated kidney differs in its two parts.

The clinical picture of the disease comprises the following symptoms: lumbar pain, fever, dysuria, tenderness in the lumbar region and hypochondrium, positive Pasternatskii's sign, arterial hypertension, pyuria, haematuria, proteinuria and bacteriuria. Pain and pyuria were the most commonly encountered symptoms in our patients. Urine culture produced growths of coli bacillus or a variety of coccal flora. The disease is usually chronic.

The peculiarities of the diagnosis in such cases are associated with the need to recognize not only the disease, but also the developmental anomaly of the kidney. Cystoscopy and excretory urography help to make an exact diagnosis of the anomaly if the function of the whole kidney is preserved even in its partial duplication. For exact diagnosis of the disease and for the diagnosis of the renal anomaly in cases of poor functioning of the whole kidney or of a part of it, retrograde pyelography is necessary, and in more difficult cases pneumoretroperitoneum or renal angiography is essential. The findings of urokymography in these cases point to marked disorders of the urodynamics of the upper urinary tract manifested in the absence of contractions of the renal pelvis and ureter or the appearance of arrhythmic and often weakened contractions. In many cases we noted the slow alternation of the contraction waves and their retrodirection, as well as lateral displacement and deformity of the ureters during contraction.

This disease is often caused by vesicoureteral refluxes which, according to Ambrose and Nicolson (1964), are noted in 50 per cent of children with duplicated kidneys. It is our experience that the reflux often occurs into the main ureter of the double kidney, less frequently into both ureters, and even less frequently into the accessory ureter. Vesicoureteral refluxes are usually attended by pyelonephritis and hydroureteronephrosis.

Hydronephrosis. In patients with double kidneys this disease can be caused by impaired tonus of the urinary tract, high origin of the ureter and its compression by accessory vessels, and constriction of the pyeloureteral segment. The most commonly encountered causes of hydronephrosis in this developmental anomaly are ectopia of the ureteral orifice, ureterocele, and vesicoureteral and interureteral refluxes.

The whole duplicated kidney or one of its parts may be involved in hydronephrosis. Goyawna and Greene (1945) contend that

the lower part of the kidney suffers more frequently, but according to Ostropolskaya (1968) affection of the upper part is more common. In our experience hydronephrosis occurred more often in the lower part of a duplicated kidney, less frequently in the whole kidney, and very rarely in its upper part.

This disease of a duplicated kidney follows a chronic course. Its duration in our cases ranged between one and twenty years. The clinical picture has no special features and is characterized by the following symptoms: pain and tenderness in the lumbar region and hypochondrium, positive Pasternatskii's sign, a palpable kidney, dysuria, fever, arterial hypertension, proteinuria, haematuria, pyuria and bacteriuria. In most cases hydronephrosis of the duplicated kidney was complicated by pyelonephritis and sometimes by nephrolithiasis.

The diagnosis of the anomaly and the disease is usually based on the findings of cystoscopy and chromocystoscopy, during which an accessory ureteral orifice may be detected, and of excretory urography which, in cases of preserved function of the kidney, demonstrates duplicated urinary tracts and dilatation of the pelvis and ureter on the radiographs. In cases of incomplete duplication and diminished function of the kidney, duplication and dilatation of the urinary tract can be revealed by retrograde pyelography, which is sometimes conducted together with tomography or pneumoretroperitoneum. In one of our cases, with reduced renal function, the disease was revealed by means of cystography. On the radiograph the vesicoureteral reflux made it possible to see clearly both ureters and pelves of the duplicated kidney. The findings of urokymography in this group of patients indicate considerable disorders in the urodynamics of the upper urinary tract, which are displayed in disorders of rhythm and weakening of contractions, slow alternation of waves, deformity of the ureters where they come in contact, disturbances of the systole-diastole complex, lateral displacement of the ureter, etc. The methods of examination mentioned above allow an exact diagnosis of this anomaly and disease to be made in all cases.

Nephrolithiasis. It is our experience that stones are usually located in the lower part of a duplicated kidney and less frequently in both parts or in the upper part.

The clinical picture of the disease in patients with a duplicated kidney is characterized, as in cases with a normally developed kidney, by lumbar pain that is dull or has the character of attacks of renal colic, disorders of micturition, discharge of stones, positive Pasternatskii's sign during exacerbation of the disease, a palpable kidney, arterial hypertension, fever, pyuria, haematuria and proteinuria. In our patients pain, positive Pasternatskii's sign, haematuria, pyuria and proteinuria

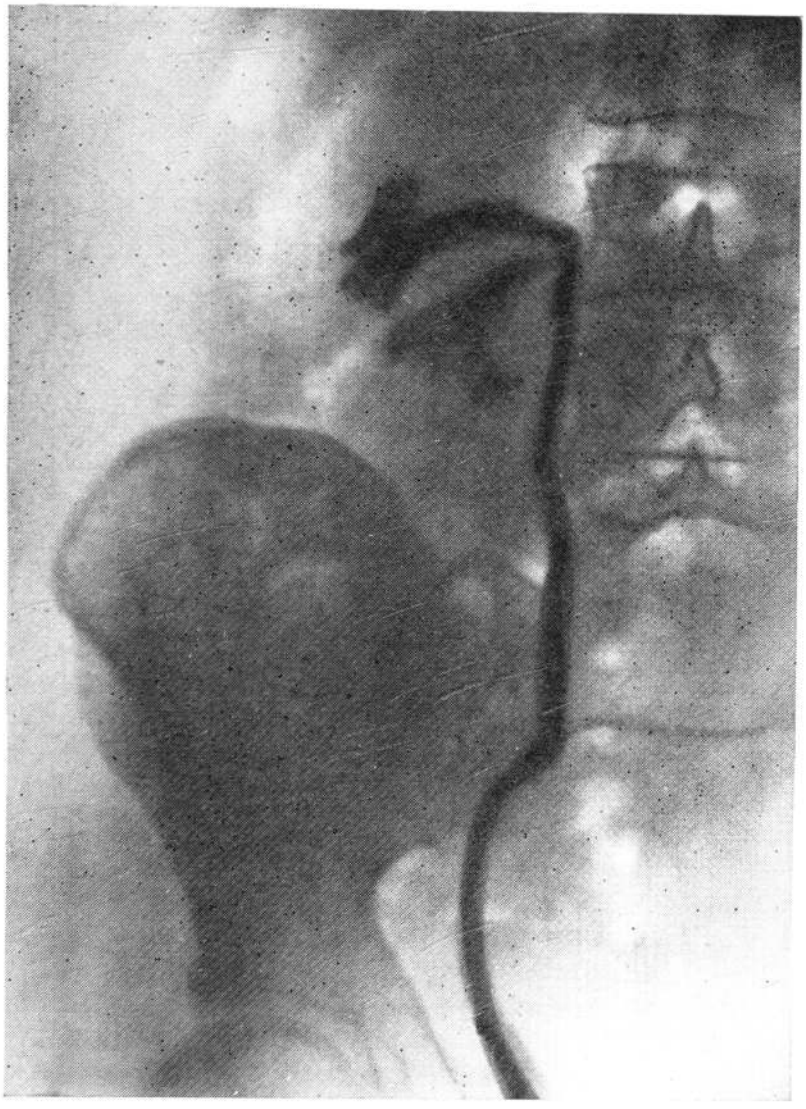


Fig. 101. Case M. Duplication of the right kidney, nephroptosis. Retrograde pyelography

were the most common symptoms. In most cases nephrolithiasis of duplicated kidneys was complicated by pyelonephritis and, in some patients, by hydronephrosis or pyonephrosis.

The diagnosis of this anomaly and disease was based on the findings of cystoscopy and X-ray examination. In some cases the anomaly was recognized because accessory ureters were found in the urinary bladder, after which the diagnosis of the anomaly was confirmed and the disease established from the findings of X-ray examination (scout radiographs and excretory urography). In other cases the diagnosis of the anomaly and disease was made during X-ray examination for nephrolithiasis.

In some cases scout radiographs lead us to suspect the presence of stones in a duplicated kidney because of the shape and position of the concrements, which resembled cavities in the duplicated kidney. The degree of duplication in such kidneys was precisely determined by cystoscopy, excretory urography or retrograde pyelography.

Excretory urography revealed a duplicated kidney in most cases with good functioning of the whole anomalous kidney. In cases of lowered function of such kidney or part of it, the diagnosis was made by retrograde pyelography. In this group also the findings of urokymography testified to disorders of the urodynamics of the upper urinary tract of duplicated kidneys. The radiographs of our patients often showed atonia, retrodirected and arrhythmic contractions, presystolic ureteral contractions, lateral displacement and deformities of the ureters at the site of their contact.

Nephroptosis. As in cases with normally developed kidneys, the clinical picture of this disease in a duplicated kidney is made up of the symptoms of pain associated with physical load on the palpated kidney or part of it, positive Pasternatskii's sign during exacerbation of the disease, disorders of micturition, haematuria, pyuria, proteinuria, arterial hypertension, etc. The disease is chronic. Over 50 per cent of such patients also suffer from pyelonephritis.

The diagnosis of the anomaly and nephroptosis, like the diagnosis of the groups of diseases described above, was based on the findings of cystoscopy and excretory urography; to rule out other diseases, it was sometimes necessary to resort to retrograde pyelography. These methods of X-ray examination yield sufficient information for making a correct diagnosis of the anomaly and the disease. The radiographs show the low position of the pelves and ureters (Fig. 101).

In our experience urokymography in this group of patients showed considerable dyskinetic changes in the upper urinary tract, the findings being the same as were described in the groups discussed above.

Tuberculosis. The clinical picture of this disease in a duplicated kidney is usually composed of the following symptoms: dull or periodically sharper pain in the lumbar region, disorders of micturition, positive Pasternatskii's sign, proteinuria, pyuria, haematuria, the presence of *Mycobacterium tuberculosis* in the urine, focal hyperaemia of the mucous membrane of the bladder with specific tubercles and ulcers on it, etc. The disease follows a chronic course.

A study undertaken by Kalenkovich (1970), a worker in our clinic, showed that the specific tuberculous process may sepa-

rately affect the upper or lower part of the duplicated kidney. Most frequently, however, both parts are involved, somewhat less frequently the lower part, and very rarely the upper part.

The diagnosis of tuberculosis of a duplicated kidney is also based on the findings of chromocystoscopy and X-ray examination and on the determination of pyuria and identification of *Mycobacterium tuberculosis* in the urine. The latter, however, like specific tubercles on the mucous membrane of the bladder, are encountered very rarely. Usually the diagnosis has to be made on the grounds of accessory cavities or deformity of the renal pyelo-calyceal system, which are shown by retrograde pyelography.

Radiographs in tuberculosis of duplicated kidneys may cause diagnostic errors which can be grouped as follows: (1) X-ray signs of a duplicated kidney are mistaken for those of the disease; (2) a correct diagnosis of the duplicated kidney is made but the X-ray signs of the disease are mistaken for symptoms of the anomaly; (3) a correct diagnosis of the disease is made but the anomaly is not recognized.

When only the lower pelvis and its calyces are seen on the radiographs, the flattened upper calyx of the duplicated kidney is sometimes taken for compression of the calyx in disconnected cavern. If only the upper rudimentary pelvis is visible, it is sometimes interpreted as a specific proliferative process in the kidney. A branching pattern of the calyces of a duplicated kidney is taken as a sign of tuberculosis of a duplicated kidney. When the radiograph only shows one pelvis and splitting of the ureter, the diagnosis is very difficult and is sometimes made only during surgery. The following case is given in illustration.

I., a 29-year-old man, was admitted to the clinic on May 27, 1964 with diagnosis of polycavernous tuberculosis of the right kidney. He had received the antibacterial treatment for one year. Urinalysis showed pyuria; *Mycobacterium tuberculosis* had been found in the past. Excretory urography showed that the right kidney did not function; there was a normal pelvis on the left. Retrograde pyelography demonstrated destructive changes in the right kidney. Right nephroureterectomy was performed. Duplication of the kidney and splitting of its ureter in the upper third were found during operation. Microscopic examination revealed polycavernous tuberculosis of the lower part of the kidney.

In this case the contrast medium in retrograde pyelography only entered the lower pelvis and the kidney did not function. The diagnosis could evidently have been made before operation by means of renal angiography. Attention should be drawn to the absence of specific changes in the upper part of the kidney in cases of polycavernous lesion in the lower part.

Tumours. Occasional cases of tumour in duplicated kidneys

have been described in the literature. There are reports on hypernephroma of a duplicated kidney (Vdovin, 1962), papillary carcinoma (Shackman and Billimoria, 1942), papillary cystadenoma (Braasch and Scholl, 1922), reticulum-cell sarcoma (Gavryliv, 1966), liposarcoma (Mukhin, 1957), leiomyosarcoma (Higbee and Atkins, 1954), leiomyoma (Connseller and Monviile, 1936), and cavernous haemangioma (Ovnatanyan, 1951).

We have had six patients with hypernephroma of duplicated kidneys. In most cases the tumour was in the lower part of the kidney, in one case in the upper part, and in another case the whole kidney was affected.

The clinical picture of this disease in duplicated kidneys has no specific features. The diagnosis is based on the clinical findings and the results of cystoscopy, excretory urography or retrograde pyelography. The discovery of an accessory ureteral orifice in the bladder during cystoscopy and a duplicated urinary tract during excretory urography helps to make the correct diagnosis of the anomaly. A filling defect in the pelvis or calyces demonstrated by excretory urography or retrograde pyelography verifies the diagnosis of a new growth in such a kidney. In one of our cases we were able to make this diagnosis by combining tomography with excretory urography (Fig. 102).

Pneumoretroperitoneum and renal angiography help in revealing the tumour in difficult cases.

TREATMENT OF DUPLICATED KIDNEYS AND THEIR DISEASES

A duplicated kidney which has no clinical manifestations and has no disease does not require treatment. Non-operative treatment, consisting of medication with spasmolytics and catheterization of the renal pelves, aimed at improving the flow of urine from the upper urinary tract and relieving dyskinesia, is applied in cases when it causes pain. According to the data in the literature and our observations such treatment brings only temporary improvement. Renal decapsulation (Mikhelson, 1929) or removal of the upper part of the kidney (Gutierrez, 1944; Williams and deBacker, 1962) performed by some surgeons is also ineffective.

It is our experience that marked disorders of the urodynamics of the upper urinary tract due to dyskinesia can be corrected by organ-preserving operations in which one of ureters is removed and high-level pyeloureterostomy is performed. This manipulation removes the cause of the pain and prevents the development of diseases associated with poor drainage of the pelves. We performed such operations on three patients and they proved successful. One of these cases is described below.

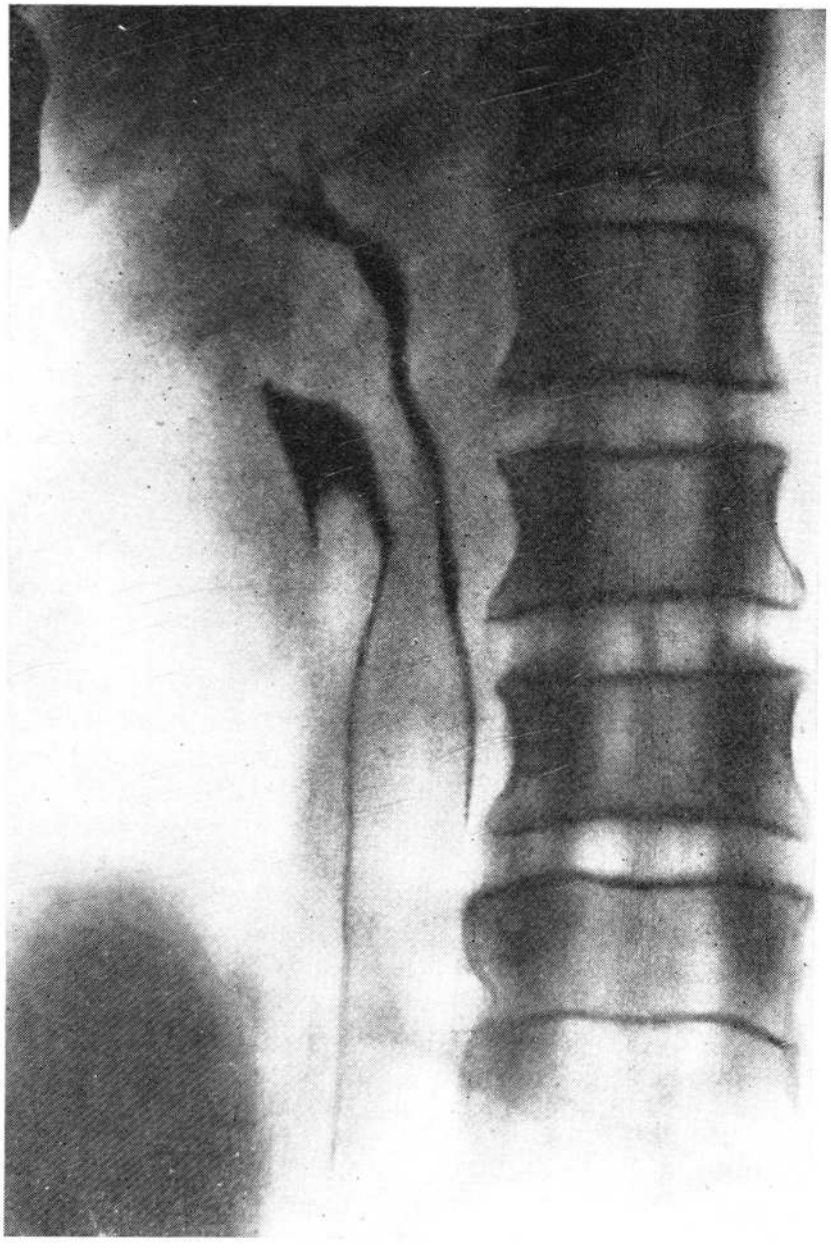


Fig. 102. Case K. Hypernephroma of the lower half of the right duplicated kidney. Excretory urography and tomography

S., a 27-year-old woman, was admitted to the clinic on September 14, 1970. She complained of attacks of pain in the left lumbar region between which there were periods of dull pain in the same area. She had been ill five years. Treatment with spasmolytics and catheterization of the pelvis brought temporary relief. Palpation elicited tenderness in the left hypochondrium. Pasternatskii's sign was positive on the left. Blood and urine tests showed normal values. Chromocystoscopy: one ureteral orifice was seen on the right and two orifices on the left; indigo carmine was discharged from

the right ureteral orifice in the third minute and from the left orifices in the fifth minute. A scout radiograph did not show any shadows of concrements. Excretory urography showed satisfactory functioning of both kidneys, and two pelves and two ureters on the left, with the lower pelvis dilated slightly. Retrograde pyelography demonstrated two pelves and two ureters on the left, the lower pelvis being slightly dilated.

Urokymography showed arrhythmic contractions, presystolic contractions, and lateral displacement of both ureters and deformity of their lumina at the sites of contact. No signs of a disease of the duplicated kidney were revealed. Dyskinesia of the upper urinary tract of the left duplicated kidney was diagnosed. An operation was performed during which the upper ureter was divided at the level of the lower pelvis and removed down to its orifice in the bladder. Its end was anastomosed with the side of the lower pelvis. The postoperative period was uneventful. The patient has a follow-up period of three years. She has no complaints, and laboratory tests and instrumental and X-ray examination reveal no abnormalities. Excretory urography shows adequate functioning of the left kidney, both pelves of normal size, the site of the anastomosis, and the ureter distal to the anastomosis. The patient recovered.

The success of such operations in dyskinesia of the upper urinary tract of duplicated kidneys allows us to recommend them for such cases.

Blind termination of the ureter in the distal or proximal segment is managed by surgery. The character of the operation is determined by the functional and morphological changes in the kidney and its urinary tract. Heminephrectomy and ureterectomy are indicated in cases of blind termination of the distal segment of the ureter and death of the renal segment; nephrectomy should be undertaken if the whole kidney is not functioning. Good functional and morphological condition of such a kidney allowed Ermolin (1964) to perform pyeloureterostomy successfully in one of his cases. A ureter terminating blindly in the proximal segment is resected down to its orifice in the bladder or the site of the splitting.

Ureterocele of the ureter of a duplicated kidney is treated surgically. The operation should be undertaken as early as possible so as to prevent marked functional and morphological changes in the kidney. Here also the surgeon's tactics are determined by the condition of the kidney and urinary tract. Considerable morphological and functional abnormalities in the renal segment indicate the need for heminephrectomy and ureterectomy; nephroureterectomy is carried out in cases of destruction of the whole kidney. A small ureterocele can be incised endovesically; a large ureterocele is resected and an antireflux operation performed. After ureterocele resection, Gregoir (1958) created lateral cystoureteral anastomosis, whereas Ermolin (1964) performed Boari's operation. In one of his cases Stefan (1966) excised the ureterocele with the corresponding ureter and carried out pyeloureterostomy.

Ectopia of the orifice of a ureter of a duplicated kidney usually

calls for operative treatment. Surgery is not needed only for patients with suprasphincteral ectopia of the ureteral orifice with no impairment of the passage of urine from the upper urinary tract. But these people should be kept under constant surveillance because this developmental anomaly frequently causes diseases.

Surgery for suprasphincteral and extrasphincteral ectopia of the ureteral orifices of a duplicated kidney is indicated when there are morphological or functional changes in the kidney and ureter due to disturbed urine drainage, or incontinence of urine in patients with ectopia of the ureteral orifice of the second group. The character of the operation in such cases depends on the functional and morphological changes in the kidney and upper urinary tract and the surgeon's skill. We operated on 16 of our 17 patients; uretero-ureterostomy was performed in two patients, ureteroneopyelostomy in four, cysto-ureterostomy in one, the accessory ureter was ligated in one patient, heminephrectomy was performed in seven and secondary nephrectomy in one patient. In uretero-ureterostomy and ureteroneopyelostomy the ureter with the ectopic orifice was resected during the operation. Heminephrectomy was made much easier with the use of the mechanical tantalum suture which was applied by means of the УКЛ-60 stitcher. Secondary nephrectomy had to be resorted to because of a poor result following cysto-ureterostomy.

In ectopia of the ureteral orifice the ureter and the corresponding renal segment are often greatly changed and so heminephrectomy, with resection of the involved part of the kidney and its ureter, has to be undertaken in many such cases. We believe that heminephrectomy should be carried out only when there are serious morphological changes associated with hydronephrosis or pyelonephritis in part of the kidney, and restoration of the function of this part of the kidney cannot be expected. We give one of our cases in illustration.

B., a 5-year-old girl, was admitted to the clinic on September 24, 1968. From birth she had suffered from urinary incontinence although the act of voluntary micturition was preserved. On inspection attention was drawn to maceration of the skin on the thighs, a characteristic sign of ectopia of the ureteral orifice. When indigo carmine was administered into the bladder, a tampon in the perineum was stained with a clear fluid. Blood and urine tests showed normal values. No abnormalities were seen on a scout radiograph. Excretory urography showed a clear image of one pelvis on the right and another on the left; the left pelvis was situated low in relation to the image of the kidney. A thick rubber catheter was left in the ureter for 24 hours, which caused pain in the left lumbar region and fever. The following diagnosis was made: ectopia of the orifice of an accessory ureter of a left duplicated kidney into the anterior urethra. Operation was performed during which a duplicated kidney was found; its upper part was flabby with cicatricial retractions on the surface. The ureter of the upper part was dilated and tortuous. Each renal segment had a separated vascular bundle. The vessels

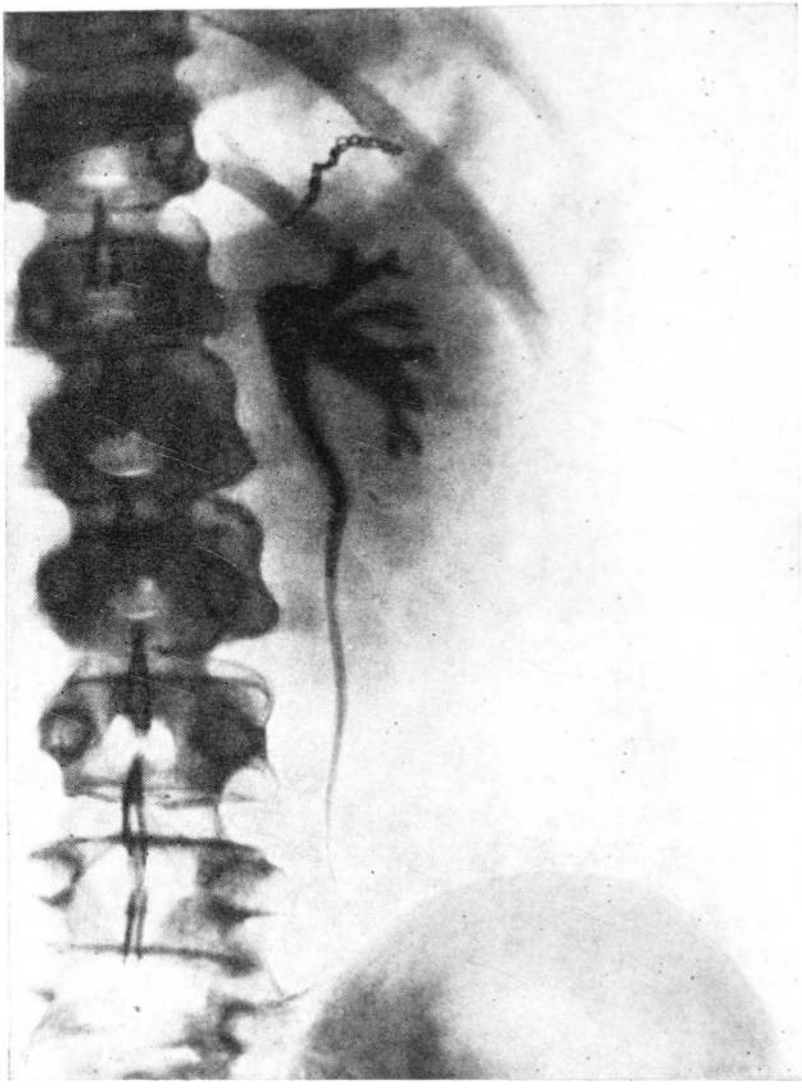


Fig. 103. Case B. After an operation for heminephrectomy and resection of the accessory ureter with the ectopic orifice. Excretory urography. Good functioning of the kidney which had been operated on. A chain of tantalum staples is seen in the upper pole of the kidney

of the upper part of the kidney were ligated. The УКЛ-60 stitcher was applied at the level of the separating groove of the anomalous kidney, the organ stitched through with tantalum sutures, and the upper segment cut off. The ureter was exposed to the level of the bladder, ligated, and resected. The postoperative period was uneventful.

Examination of the removed specimen revealed that many of the renal glomeruli were sclerosed and their capsules thickened; serous fluid was seen in some of them and there was diffuse round-cell infiltration around the glomeruli.

The patient's follow-up period is now four years. She has no complaints and controls urination adequately; blood and urine tests yield normal values.

Excretory urography shows good functioning of both kidneys. A chain of tantalum staples is seen above the left kidney on the radiograph (Fig. 103).

This case shows that induction of ureteral obturation to determine the side of the ectopia causes attacks of pyelonephritis and is therefore called for, particularly when operation with more preservation of the organ may be undertaken. The considerable morphological and functional changes in the accessory segment of the duplicated kidney in the case discussed indicated the need for removal of part of the kidney and the accessory ureter, which made the treatment successful.

Cases of high ligation of the ureter when there are marked functional and morphological changes in the segment, with ectopia of the ureteral orifice, are sometimes described.

Ligation of the ureter at the pelvis leads to atrophy of the corresponding renal segment. Bundikov (1950) and Pekarskaya (1955) used this method in the management of ectopia of the orifice of the accessory ureter of a duplicated kidney. It has even been used in hydronephrosis of the involved part of the kidney (Mikhelson, 1939), while some authors consider it indicated in the absence of infection and diminished function of this renal segment (Gektin, 1935). Hepler (1947) prefers ureteral ligation in cases with a solitary kidney.

Kawasoje's ureteral ligation technique is the most reliable; the ureter is tied with a double knot and the knots are ligated. Braude's method, tying off the ureter with two simple knots, is also a reliable procedure. This operation, however, is sometimes complicated by hydronephrosis or seepage of urine under the renal capsule (Epshtein, 1950). Chronic pyelonephritis developed in the remaining part of the kidney after this operation in one of our cases. That is why we cannot recommend it as the operation of choice.

Good functional and morphological condition of a renal segment with an ectopic ureteral orifice makes it possible to perform uretero-ureterostomy or ureteroneopyelostomy. More and more such operations are described in the literature (Filippova, 1940; Sandegard, 1958). These are the best organ-preserving operations. Six of our patients with ectopia of the orifice of the ureter of a duplicated kidney underwent such operations with good remote results. One of these cases is given in illustration.

D., an 18-year-old girl, was admitted to the clinic on February 9, 1961 complaining of urinary incontinence from birth. The act of voluntary micturition was preserved. Drops of a clear fluid were discharged from the external urethral opening. The colour of the fluid did not change after indigo carmine was introduced into the bladder. Blood and urine tests showed normal values. On cystoscopy two ureteral orifices were found on the left and one orifice on the right side. An accessory ureteral orifice was discovered beyond

the external urethral opening during urethroscopy. No shadows of concrements could be seen on a scout radiograph. An excretory urogram showed good functioning of the kidneys and their bilateral duplication. Complete duplication of the left kidney and downward displacement of the right pelvis were shown on a retrograde pyelogram.

Diagnosis: complete bilateral kidney duplication, ectopia of the orifice of the right ureter into the urethra. Operation was carried out on March 9, 1961. A right duplicated kidney was found, with no separating groove. Two ureters were exposed at the lower pole of the kidney; an elastic catheter had been introduced into the main one previously. The accessory ureter was divided at the lower pole of the kidney and its distal end resected down to the level of the bladder. The proximal end was ligated twice and a side-to-side anastomosis 3 cm long was established between both ureters by means of a single-row interrupted suture. The postoperative period was uneventful.

The duration of the follow-up period is now 10 years. There are no complaints. Blood and urine tests yield normal values. The woman gave birth to a normal baby two years after the operation. During chromocystoscopy, indigo carmine was discharged in a good stream from all three orifices in the fourth minute. Excretory urography showed good functioning of both kidneys; good images were obtained of both right pelves, the ureters, and the anastomosis; the upper urinary tract was not dilated.

This case confirms the expediency of establishing interureteral anastomosis in the management of an ectopic ureteral orifice when the duplicated kidney functions adequately and there is no infection in the urinary tract.

If the main pelvis is well developed it is more advisable to perform ureteroneopyelostomy, because the procedure is simpler and creates a higher splitting which rules out the later development of dyskinesia of the adjacent ureter. In bilateral ectopic ureteral orifices similar plastic operations have to be carried out on each side in some cases.

In one of our cases ureteral ectopia was at first recognized on only one side because one of the renal segments with an ectopic orifice functioned poorly. Since the operation was unsuccessful, an ectopic ureteral orifice on the other side was suspected and the diagnosis was verified. Good functioning of one of the renal segments with an ectopic ureter allowed us to perform successfully ureteroneopyelostomy. The segment of the contralateral kidney with the ectopic ureter was so changed that it had to be resected.

In children the urinary tract has a small lumen and there is more danger than in adults of strictures forming in interureteral anastomosis. That is why ureteroneopyelostomy is preferred over uretero-ureterostomy in such cases.

When the renal segment with the ectopic ureter and the ureter function adequately some urologists transplant the ureter into the bladder (Mikhelson, 1929; Salturin, 1940). The frequent poor results of this operation, however, have prompted other urologists (Blundon and Lane, 1960) to disapprove of it. This operation is evidently

necessary in ectopia of the orifice of the common ureter or in ectopia of the orifices of both ureters of one kidney. Gasparyan (1935) contends that it should be undertaken in ectopia of the orifice of a common ureter despite the risk of repeated nephrectomy.

Among the complications involved in this operation constriction of the anastomosis, vesico-ureteral reflux, and inflammation in the kidney are often described (Epshtein, 1950). Schlachetzka (1966) claims that however well the operation is performed normal passage of urine is always impaired after ureteral implantation.

Transplantation of an ectopic common ureter into the bladder was conducted in one of our patients. But because of obliteration of the new anastomosis in the bladder and atrophy of the kidney, secondary nephrectomy had to be performed. We find this operation unwarranted also because it does not correct the dyskinesia in the ureters and pelves. Even if its immediate results are satisfactory pain and diseases develop later. For this reason we believe that in heminephrectomy and anastomosis of the upper urinary tract it is necessary to resect the whole ectopic ureter. Inflammation develops sometimes in the stump of the remaining ureter due to impaired drainage and, in addition, contractions of the remaining part of the ureter cause disorders in the contractions of the main ureter and hinder the flow of urine from it. We encountered this during examination of such patients by urokymography.

Hydronephrosis of a duplicated kidney is managed by surgery. Judging from the data in the literature, nephrectomy or heminephrectomy is the preferred operation in such cases and most authors name the latter as the operation of choice (Ermolin, 1964; Ostropolskaya, 1968; Bairov, 1969).

It is our experience that non-operative treatment of this disease of duplicated kidneys (catheterization of the renal pelves, treatment with spasmolytics, anti-inflammatory therapy) produces temporary improvement. The disease continues to progress steadily, which is why such treatment can be recommended only at the onset of the disease and for a short time. If it proves unsuccessful, it is not advisable to temporize until considerable morphological disorders develop in the kidney and ureter and their function is lost. An organ-preserving operation must be undertaken in time.

Our clinic possesses information on 41 operations conducted in hydronephrosis of duplicated kidneys: six nephrectomies, 18 heminephrectomies, four ureteroneopyelostomies, three pyeloureterostomies, two uretero-ureterostomies, two Y ureteroneopyelostomies, one Y uretero-ureterostomy, and five operations with plastics of the renal pelvis by various methods. The success of

the treatment in most cases allows us to give some recommendations with regard to this problem.

We consider nephrectomy advisable in cases of infected hydronephrosis of both halves of the kidney and loss of function, hydronephrosis of half of the kidney and pyelonephritis of the other half with loss of function of the whole kidney, and hydronephrosis of one segment of the kidney and atrophy of the other segment also with loss of function of the whole kidney. Heminephrectomy may be undertaken when one segment of the duplicated kidney is involved in hydronephrosis while the other segment functions adequately. According to our experience, the renal segment remaining after such an operation may function well and not trouble the patient. The following case from our practice is given in illustration.

D., a 51-year-old man, was admitted to the clinic on February 29, 1964. He complained of difficult micturition, periodical dysuria, and dull pain in the left lumbar region. He had a urethral stricture after fracture of the pelvic bones in 1944 and was subjected to bougienage at regular intervals. The lower half of the left kidney was palpated. Blood tests showed normal values. Urinalysis: alkaline reaction; specific gravity 1014; protein 1.65 pro mille; leucocytes covering the entire field of vision, 30-40 erythrocytes in field of vision. Urine culture produced growth of coli bacillus. Cystoscopy revealed three ureteral orifices in the bladder, two of them on the left. No shadows of concrements were found on a scout radiograph. Excretory urography showed the shadow of a normal pelvis on the right and the shadow of a rudimentary pelvis on the left against the background of a shadow of an elongated kidney and the shadow of the pelvis in the upper renal pole. Reflux into the ureter, which was continuous with the shadow of a large pelvis, was seen on the cystogram. Retrograde pyelography disclosed a duplicated left kidney with a rudimentary upper pelvis and a very large lower pelvis. Hydronephrosis of the lower part of a duplicated kidney with vesico-ureteral reflux into its ureter was diagnosed. Heminephrectomy of the lower part of the duplicated kidney was carried out. The post-operative period was uneventful. Control examination eight years later showed that there were no complaints of a kidney disease, and blood and urine tests showed normal values; good functioning of the upper part of the left duplicated kidney was demonstrated by excretory urography.

The favourable results of heminephrectomy in diseases of duplicated kidneys allowed some authors to recommend its wide use in cases of isolated affections of parts of such anomalous kidneys (Ostropolskaya, 1968). Our experience shows, however, that this operation should not be undertaken widely in such cases, because when the function of both renal segments is maintained it often proves possible to preserve the whole kidney by ureteroneopyelostomy. We recommend heminephrectomy in cases of gross destruction of the renal tissue and impaired function of a part of the kidney, when the outcome of a plastic operation is doubtful.

In cases of duplicated kidney, heminephrectomy is much easier

if a mechanical tantalum suture, applied by means of the УКЛ-60 stitcher, is used. The technique of such an operation is quite easy: after exposure of the duplicated kidney, separation of the vascular bundle, and ligation of the vessels the blades of the stitcher are placed on the separating groove between the parts of the anomalous kidney and gradually drawn together. The kidney is then stitched through with tantalum staples, the part to be resected is cut off, and the stitcher removed. After such operations we encountered neither haemorrhage nor any of those severe complications which are often described in heminephrectomy, namely, progressive pyelonephritis of the remaining part of the kidney, apostematous nephritis, thrombosis of the renal vessels, disturbance in the passage of urine, urinary fistulas (Hamm and Finkelstein, 1959; Khomenko, 1967). These complications make it necessary to perform another nephrectomy.

In mild or moderate grade I-II hydronephrosis, when the whole kidney still functions, it is expedient to perform plastic procedures of the uretero-ureterostomy or ureteroneopyelostomy type.

These operations are undertaken in cases of stenosis of the prepelvic ureteral segment (Fowler, 1959), constriction of the ureter at the site of its splitting or in the middle part (Gregoir, 1958), and in megaloureter (Smolyak and Svidler, 1965).

Our observation of twelve patients subjected to such operations (ureteroneopyelostomy in four, pyeloureterostomy in three, Y ureteroneopyelostomy in two, uretero-ureterostomy in two, and Y uretero-ureterostomy in one patient) showed good remote results in most with follow-up periods of two to seven years. In only one patient, with a lateral Y uretero-ureterostomy established at a low level, was the result of the operation poor, due to the progressive pyelonephritis. We think that the operation was not indicated in this case because the function of the renal segment was considerably diminished before surgery and there was severe chronic pyelonephritis. Heminephrectomy should have been performed instead. There was no stenosis of the anastomosis after the operation, but the function of the kidney deteriorated progressively.

It should be noted that in the other six cases chronic pyelonephritis was milder and with the creation of good drainage of the urinary tract the condition ended in clinical recovery, since in these patients the remote results were good. For this reason we believe that when the kidney is functioning adequately and there is no marked destruction of the renal tissue (determined by X-ray examination), pyelonephritis is not a contraindication for performing these operations. The following case from our practice serves as an illustration.

Fig. 104. Case K. Duplicated left kidney with ureteral splitting. Hydronephrotic transformation of the lower half of the kidney due to stricture of the ureter. Retrograde pyelography

K., a 25-year-old woman, was admitted to the clinic on October 23, 1964. She complained of pain in the left abdomen and frequent and painful micturition. The pain often intensified and took the character of renal colic. She had been ill for three years. Pasternatskii's sign was positive on the left. Blood tests showed no abnormalities. Urinalysis: acid reaction; specific gravity 1017; protein 0.66 pro mille; 30-40 leucocytes in field of vision. Urine culture produced enterococcal growth. Urine from the right kidney contained three or four leucocytes in field of vision. With chromocystoscopy a ureteral orifice was seen on each side of the bladder, indigo carmine being discharged in the fourth minute from both. Excretory urography indicated good functioning of both kidneys and showed one pelvis on the right and two pelves on the left. Retrograde pyelography on the left showed splitting of the ureter into two branches in the upper third and constriction of the prepelvic segment of the lower pelvis (Fig. 104).

Fig. 105. The same patient as in Fig. 104. Excretory urography made six years after operation. Good functioning of the kidney

A diagnosis of hydronephrosis of the lower part of the duplicated kidney with splitting of the ureter in the upper third and pyelonephritis was made. Stricture of the prepelvic segment of the main ureter was found to be the cause of hydronephrosis. Surgery was performed. The duplicated kidney was exposed and a hard constriction was found in the prepelvic part of the lower ureter. The ureter was divided above the level of the constriction and excised down to the site of the splitting. The lower pelvis was sutured into an incision made in the upper ureter (end to side). Intubation of the anastomosis and drainage of the kidney were not applied. The postoperative period was uneventful. The patient recovered. The follow-up period was seven years, during which time the patient had no complaints, pain did not trouble her, and she had no attacks of pyelonephritis. The kidney which had been operated on functions adequately. Excretory urography (Fig. 105) showed two pelves, the anastomosis, and ureter; the urinary tract was not dilated. Blood and urine tests yielded normal values.

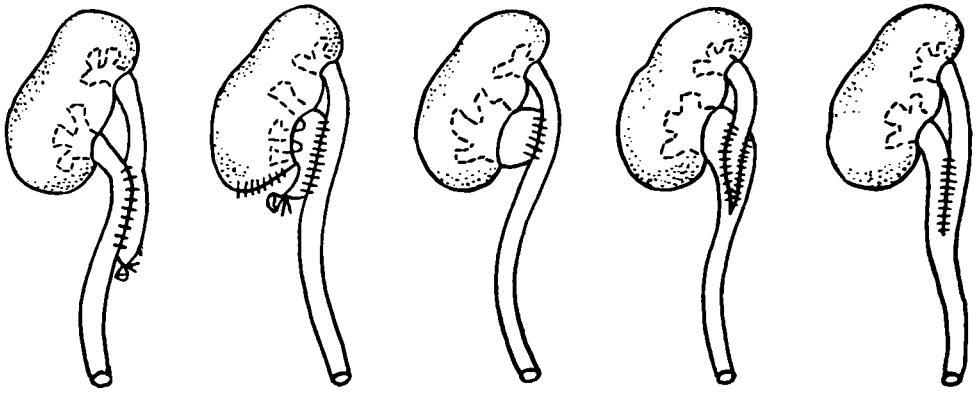


Fig. 106. Variants of uretero-ureterostomy and pyeloureterostomy

Pyeloureterostomy was conducted in this case (the lower pelvis was stitched to the accessory ureter). An adequate flow of urine from both pelves was conducive to the abatement of the inflammatory process and recovery. There is no doubt that anti-inflammatory therapy applied in the first postoperative years to prevent pyelonephritis contributed to this.

The techniques of uretero-ureterostomy and pyeloureterostomy are marked by some peculiarities which are shown in Fig. 106.

The communication between the renal pelves and the ureters is made 2 or 3 cm long with interrupted catgut sutures applied in one row. To avoid constriction of a pyeloureteral anastomosis, more of the pelvic wall and less of the ureteral wall should be taken with the suture; in the case of uretero-ureterostomy the sutures are applied close to the edge on both sides. Care should be taken not to place the sutures close to each other at the corners of the anastomosis because it is here that the stenosis occurs.

We undertook pyeloureterostomy in stenosis of the prepelvic segment of the main ureter. The main ureter was excised with the constriction down to the level of the splitting and the end of the lower pelvis stitched into an opening in the upper ureter (end to side). In another case such an operation was a component of combined surgery on a horseshoe kidney. After the renal isthmus had been resected, the main ureter was excised and the accessory ureter was anastomosed side to side with the lower pelvis (the main ureter was ligated below the main pelvis). We were thus able to remove the ureter in vesico-ureteral reflux and pyelonephritis, removing the cause of the disease.

In a case of constriction of both ureters of a duplicated kidney (the main one in the prepelvic segment and the accessory ureter at the site of the splitting) we corrected the constriction of one of them at the expense of the other.

Because of the originality of this plastic operation on the kidney we cite here the case record.

R., a 35-year-old woman, was admitted to the clinic on October 10, 1967. Urological examination revealed duplication of the right kidney with splitting of the ureter in the middle third, hydronephrosis of the lower part due to stenosis of the prepelvic segment of the main ureter, ureterohydronephrosis of the upper part of the kidney as a consequence of stenosis of the upper ureter at its origin from the common ureter, and stones and pyelonephritis in the contralateral kidney. The function of both kidneys was preserved. Pain and hydronephrosis developing in both segments of the duplicated kidney were indications for surgery, but the disease (nephrolithiasis and pyelonephritis) of the contralateral kidney allowed only an organ-preserving operation.

Surgery was performed on the left duplicated kidney. The upper ureter was divided obliquely a little below the level of the constriction of the second ureter and cut longitudinally for a distance of 1 cm. The distal end of this ureter was resected down to the level of the splitting of both ureters. The lower pelvis and part of the ureter were cut through the constriction. The accessory ureter was stitched (end to side) into the defect formed in the pelvis and part of the ureter. Thus, the narrowing in the prepelvic segment of the main ureter was widened by means of the accessory ureter, forming a communication between the pelvis and ureter of both segments of the duplicated kidney at the site of the constriction.

The postoperative period was uneventful. The patient recovered. She was kept under observation for five years. The duplicated kidney did not trouble her; it functioned adequately. Tests of urine collected from this kidney showed normal values.

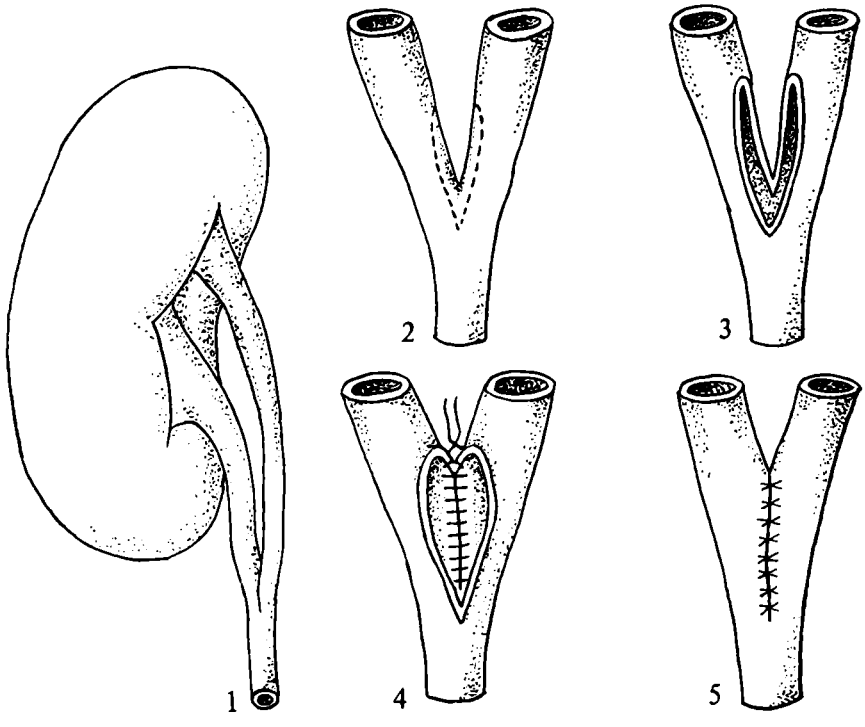


Fig. 107. Details of Y uretero-ureterostomy

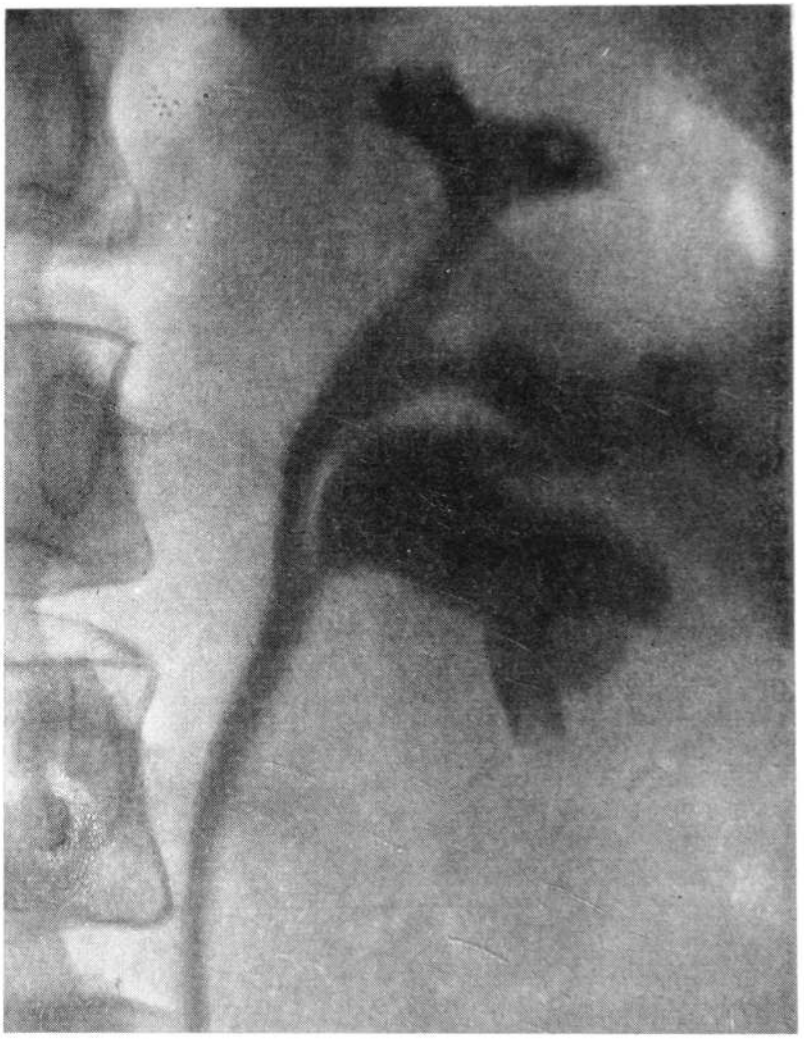


Fig. 108. Case L. Incomplete duplication of the left kidney with splitting of the ureter in the upper third, stenosis of the lower ureter at the site of the splitting and at the pelvis, hydronephrotic transformation of the lower renal segment. Retrograde pyelography

A Y uretero-ureterostomy may be performed in high splitting of the ureter. For this purpose the branches of both ureters are cut on the medial surface to the level of the splitting and through it and the edges thus formed are sutured together (Fig. 107). A still higher level of ureteral splitting is formed as a result and the obstacle to the flow of urine is removed. The following case from our practice is given in illustration.

L., a 20-year-old woman, was admitted to the clinic on February 21, 1969. She complained of continuous pain in the left lumbar region which became aggravated now and then. In the last few months she could no longer work because of the pain. The lower pole of the left kidney was palpated.

Fig. 109. The same patient as in Fig. 108 three years after an operation for uretero-ureterostomy. Good functioning of the kidney. The urinary tract is not dilated

Pasternatskii's sign was positive on the left. Blood and urine tests showed no abnormalities. Chromocystoscopy: one orifice on each side of the bladder in the region of the trigone; indigo carmine was discharged from them in the fourth minute. Excretory urography showed that both kidneys functioned adequately and that the left pelvis was dilated. Retrograde pyelography on the left side showed splitting of the ureter in the upper third, stenosis of the prepelvic segment of the ureter of the lower segment, and an enlarged shadow of the lower pelvis (Fig. 108).

The following diagnosis was made: incomplete duplication of left kidney with splitting of the ureter in the upper third, stenosis of the lower ureter at the site of the splitting and at the pelvis, hydronephrosis of the lower segment of the duplicated kidney. The continuous pain which made the patient incapable of working and hydronephrosis indicated the need for surgery. During the operation a duplicated kidney with high splitting of the ureter at the pelvis was found. The ureter of the lower segment was constricted at the site of the splitting and the lower pelvis was enlarged.

Fig. 110. Case Zh. Complete duplication of the right kidney, hydronephrotic transformation of the lower segment. Retrograde pyelography

Fig. 111. The same patient as in Fig. 110. Excretory urography performed three years after pyeloureterostomy. Good functioning of the kidney. The renal pelvis has diminished in size

An incision was made on the medial surface of the lower pelvis and part of the ureter to the site of the splitting, and then a second incision on the medial surface of the upper ureter and through the site of the ureteral splitting. The edges of the incisions of both ureters were then stitched together so that the splitting of the upper urinary tract was now above the level of the lower pelvis, i.e. Y ureteroneopyelostomy was performed (side to side). The postoperative period was uneventful. The patient recovered.

L. was kept under constant surveillance. Examination during a follow-up period of four years showed no complaints, with blood and urine tests yielding normal values. Excretory urography demonstrated adequate function of the kidney. Both pelves, the ureter and the anastomosis were clearly seen on the radiograph. Retrograde pyelography on the left side showed a clear image of the ureter, the site of the splitting, and both pelves; the urinary tract was not dilated (Fig. 109).

In Y uretero-ureterostomy, particular care should be taken so as not to make the lower part of the communication narrower by applying too many sutures which grasp large areas of the ureteral walls. This is the area of least resistance to the possible formation of a constriction.

Uretero-ureterostomy (side to side) can be performed in the management of cases of intrarenal pelves. In one of our patients with this condition we carried out uretero-ureterostomy (end to side) with good results.

We also achieved a good result when we formed an end-to-side anastomosis between the main pelvis and the accessory ureter (Figs. 110 and 111).

This case shows that end-to-side anastomosis of ureters performed by a skilled surgeon is no more hazardous than side-to-side anastomosis, from the point of view of the formation of a constriction. This is also confirmed by the fact that in our experience most operations for establishing end-to-side anastomosis were successful. We carried out in all groups 21 such operations, 14 with end-to-side anastomosis and only five by the side-to-side technique. It should be noted that when we say ureteroneopyelostomy we mean a manipulation in which the upper ureter of a duplicated kidney is sutured into the lower pelvis, whereas pyeloureterostomy is one in which the lower pelvis is sutured into the upper ureter, which in this case is preserved. We have had a poor result of the operation in the second group. Though we consider progressive pyelonephritis to be the cause of the poor outcome, since the anastomosis was not obstructed, we nonetheless cannot determine on the basis of these data the advantages of side-to-side anastomosis over the end-to-side method.

Ureteroneopyelostomy is usually accomplished by stitching the edges of the cut pelvis to the edges of the ureter. For the same purpose, however, the ureter may be invaginated into the pelvis, in which case a smaller incision is made in the pelvis. Fixation

of the ureter in the pelvis proves to be more secure by this method. We performed this operation on two of our patients. It proved to be very simple and suitable for operating on the narrow urinary tract of children, when it is difficult to stitch the edges of the pelvis to the edges of the ureter and there is greater danger of the formation of a constriction. This modification of the operation naturally cannot be used in patients with an atonic ureter, which could easily be strangulated by the muscle of the renal pelvis at the place of the anastomosis.

It should be emphasized that the anastomosis between the urinary tracts of duplicated kidneys should be formed at a high level and the whole ureter below the anastomosis should be resected. If this is not done, distorted contractions occur in the remaining ureteral segment, it empties poorly, and abnormal contractions of the main ureter are revealed at this level. In other words, the ureteral stump interferes with the normal functioning of the main ureter. Evidence of this was gained in our urokymographic study of the dynamics of the upper urinary tract after different surgical manipulations on duplicated kidneys.

Proceeding from this, it is better to perform pyelo-pyelostomy or pyeloureterostomy than uretero-ureterostomy. The first two operations rule out any contact of the ureters and associated disorders of their motor activity. To accomplish pyelo-pyelostomy, however, the pelves must be large and located close to one another, which is a rare occurrence. Pyeloureterostomy also cannot be performed if the patient has intrarenal pelves. We find it expedient to perform these manipulations in the following order: if pyelo-pyelostomy cannot be performed an attempt should be made at ureteroneopyelostomy, but if this is also not possible, then uretero-ureterostomy should be conducted. Bearing in mind the factors discussed above, we believe that in cases of duplicated kidneys the ureters should not be anastomosed with the urinary bladder.

Like Swenson and Ratner (1962), we do not consider pyelonephritis to be an absolute contraindication for an early operation. In pyelonephritis it is necessary to drain both pelves for seven to ten days. Drainage is accomplished with a perforated tube which is brought outside through the urethra. At the same time this tube serves as a prosthesis on which the anastomosis forms. Lack of intubation in one of our cases of Y uretero-ureterostomy was evidently among the causes of a poor result. The tube fell out on the second postoperative day. It should be noted in connection with this that the contracting ureter pushes the drain out and so it must be fastened to the skin on the thigh. The drain should be watched constantly and taken care of; it should be seen that the fluid flows from it and the drain should be washed

out with antiseptic solutions not only when it becomes obstructed, but also at regular intervals as a preventive measure.

The results of these plastic correcting operations in hydronephrosis have been followed up for five to ten years. Good results (no complaints, normal values in blood and urine tests, good functioning of the kidney, a decrease in the size of the pelvis and calyces) were noted in eleven patients, a poor result was found in one patient. Pain was completely relieved after Y uretero-ureterostomy, but pyuria did not cease and the function of the kidney deteriorated. We believe that the cause of this failure was progressive pyelonephritis which had lowered considerably the functional indices of this kidney before surgery and caused some technical errors during the operation (the drainage tube fell out early).

The results obtained show that these organ-preserving operations are very effective when the indications are as discussed above and when proper operative techniques and postoperative management are accomplished. They may be recommended for wide use in clinical practice but on the condition that they are conducted by surgeons sufficiently experienced in plastic operations on the upper urinary tract.

Plastic operations are rarely performed in duplication of the kidneys. The techniques and results do not differ from those in surgery on normally developed kidneys.

Ratner et al. (1961) operated on two patients with a solitary duplicated kidney replacing the deficient ureter of the contralateral duplicated kidney with a graft of the small intestine. The operation was effective in one case, but brought about no improvement in the other.

Thus, correcting operations such as pyelo-pyelostomy, ureteroneopyelostomy, pyeloureterostomy or uretero-ureterostomy as well as heminephrectomy with the removal of one of the two renal segments can be used extensively in the management of hydronephrosis of duplicated kidneys. They may be combined with plastic surgery on the upper urinary tract. Plastic operations alone are evidently insufficiently radical due to the peculiarities of the urodynamics of the upper urinary tract. In marked morphological and functional changes in both segments of the duplicated kidney nephrectomy is necessary.

Pyelonephritis of a duplicated kidney is usually treated with various drugs, just as pyelonephritis of a normally developed kidney. Judging by the late results in our cases, however, nonoperative treatment does not cure the disease completely. The disease takes a chronic course with periodical exacerbation despite treatment.

Nephrectomy is indicated in cases of marked affection of both

segments of the duplicated kidney with disturbance of its function, as well as in a unilateral process of this kind with associated high arterial hypertension.

Isolated affection of one segment of the kidney by pyelonephritis makes it possible to restrict surgery to heminephrectomy. Grieve and Thompson (1956) found such operations to be successful and attributed their efficacy to the improved urodynamics of the remaining part of the kidney.

The character of surgery in vesicoureteral reflux is also determined by the morphological and functional changes in the associated kidney. Nephrectomy is recommended in severe changes in both renal segments, and heminephrectomy when such changes are found only in one segment. When the kidney and ureter have been in fairly good condition, some urologists have resorted to antireflux-type operations. Williams and deBacker (1962) carry out ureteroneocystostomy. We consider these operations to be insufficiently radical in cases of duplicated kidney, since they do not correct the disorders of the urodynamics of duplicated urinary tracts discussed above.

These disorders may contribute to the maintenance of infection in one of the segments of the kidney even in the absence of vesicoureteral reflux. In one such case we formed an ureteropelvic anastomosis to provide an adequate flow of urine from the upper urinary tract and achieved a good result. A follow-up period of two years showed that pain and attacks of pyelonephritis were fully relieved. This operation is particularly advisable in a case of vesicoureteral reflux into one of the two ureters.

We had good results in two cases of vesicoureteral reflux and pyelonephritis of one segment of a duplicated kidney by performing ureteroneopyelostomy and pyeloureterostomy. In one of them pyeloureterostomy was a manipulation in a reconstructive operation on a horseshoe kidney with removal of the isthmus. The follow-up periods in these cases were four to five years.

The same operation is advisable in cases of vesicoureteral reflux into both ureters of a duplicated kidney, whereas an antireflux operation is conducted on one ureter.

Nephrolithiasis in cases of duplicated kidneys is treated according to the principles commonly accepted for the disease. Pyelo- and ureterolithotomy are preferred, nephrolithotomy being carried out less frequently. The results of operations in our cases have been followed up for periods of two to ten years. They were poor in 28 per cent of cases. Besides, further follow-up of patients showed that, with one exception, all had signs of pyelonephritis. The high incidence of poor results in the treatment of this disease in duplicated kidneys is usually linked with marked uro-

dynamic disorders in the upper urinary tracts of these malformed kidneys.

Considerable changes in one of the renal segments in nephrolithiasis, which are usually consequent upon calculous pyelonephritis, calculous hydronephrosis or pyonephrosis or which develop with recurrence of the disease, are indications for heminephrectomy. This operation usually produces good results in nephrolithiasis of duplicated kidneys. Five of our patients were subjected to the operation. Follow-up periods of two to four years show its results to be favourable in four patients. They have no complaints, blood and urinary tests show normal values, and no signs of pyelonephritis of the remaining renal segment are found.

Nephrectomy has to be performed in cases of nephrolithiasis when there is calculous pyelonephritis or hydronephrosis in both segments of the duplicated kidney or when heminephrectomy proves unsuccessful. We operated on three of our patients for these conditions. One of them, unfortunately, had to be treated later for pyelonephritis of the remaining kidney, the function of which was markedly diminished with arterial hypertension developing in addition. This is another proof of the expediency of performing early organ-preserving operations in nephrolithiasis.

Tuberculosis of a duplicated kidney is now usually treated by non-operative measures, but when surgery is necessary heminephrectomy is preferred.

All our patients received long-term non-operative treatment according to the commonly accepted principles of the management of tuberculosis of the urinary system; six patients underwent surgery. Nephrectomy was performed on four patients for pyonephrosis, polycavernous tuberculosis, and tuberculosis of both halves of a duplicated kidney complicated by pyelonephritis and contraction of the kidney. Examination of the removed kidneys in cases of pyonephrosis showed that in some of them the organ-preserving operations could have been carried out, since only one segment was affected. This is why in one of our patients with cavernous lesion of the upper segment of a duplicated kidney heminephrectomy proved successful. In a follow-up period of three years the postoperative results are good. Operations even more organ-preserving than resection of a renal segment can evidently be conducted on duplicated kidneys. A renal segment was successfully resected in two of our cases with a demarcated cavern in the lower segment of a duplicated kidney.

Partial resection of the lower segment with a demarcated cavern makes it possible to leave intact much of the lower segment of the duplicated kidney. Functionally this segment is more

important and aids recovery considerably. Long-term pre-and postoperative antituberculosis treatment contributes to the success of the operation.

Pathological mobility of a duplicated kidney is managed according to the commonly accepted principles for the treatment of this condition. It is our experience that non-operative treatment brings temporary relief in these cases, but pain and pyelonephritis remain. We resorted to surgery, nephropexy of the duplicated kidney, in nine patients. During the operation care must be taken to avoid stretching the vessels of the anomalous kidney, because they are often numerous and short in length. Compression of the duplicated urinary tract should also be avoided. Follow-up periods of two to five years showed that pain was totally relieved in half of the patients and was less intense in the others. Symptoms of pyelonephritis persisted in all patients after the operation, but in one patient arterial pressure, which had been high before surgery, decreased to normal levels.

Tumours of duplicated kidneys must be treated surgically, by nephrectomy. Our experience in the treatment of three patiens shows that the operation is only distinguished by the necessity of ligating the numerous vessels of the kidney. This is why care must be taken in exposure of the medial surface of the kidney. Hypernephroma of the duplicated kidney was removed in our cases. The postoperative follow-up periods are two to four years. The patients' condition is satisfactory, and there are no signs of metastatic spread.

In concluding this section on the treatment of duplicated kidneys and their diseases, it should be noted that non-operative measures bring only temporary relief in most cases because of the peculiar structure of the anomalous kidney. Operative intervention is marked by some specific features associated with the possibility of performing on this anomalous kidney such organ-preserving operations as anastomosis between the upper urinary tracts and heminephrectomy. Heminephrectomy is advisable in cases of marked morphological and functional changes in one segment, whereas anastomosis and resection of one of the two ureters are more expedient when function in both segments is preserved (the higher the anastomosis is formed, the better will be the result). We find it inadvisable to transplant the ureters of a duplicated kidney into the urinary bladder because of the marked dynamic disorders in the two ureters stretching next to each other.

As a result of the creation of the communication at a high level and removal of one of the ureters good remote results are achieved. Pain, which had been caused by dyskinesia of the upper urinary tract, is relieved. These operations produce favourable

remote results in the management of ectopic ureteral orifice and hydronephrosis, and in some cases of pyelonephritis, particularly when it is linked with dyskinesia of the upper urinary tract or vesicoureteral reflux.

The remote results of commonly performed operations in nephrolithiasis and pathological mobility of the duplicated kidney are poorer than the results of these operations on normally developed kidneys. Organ-preserving operations such as heminephrectomy and resection of the kidney may be performed in cases of tuberculosis of the kidney; they have good remote results.

SUPERNUMERARY KIDNEYS

A supernumerary kidney is a third kidney found separate from and in addition to the two independent kidneys. The parenchyma of each kidney is unconnected with the parenchyma of the other two kidneys. We believe that supernumerary kidney is the most appropriate name for this anomaly because a case of two accessory kidneys has been reported in the literature (Temelkoff, 1957). Besides the two normal kidneys there were another two independent small ones.

During the development of a **third, supernumerary, kidney,** besides the changes characteristic of the formation of a duplicated kidney, there is also splitting of the metanephrogenous tissue in the vicinity of the main and accessory ureters.

A supernumerary kidney is a very rare anomaly. It was described for the first time by Blasius as early as 1677. According to Geisinger, however, by 1937 information on only 40 cases (including that described by Blasius) could be found in the literature. The number of such cases has increased a little over the past years. Carlson (1950) published summarized statistical data concerning 51 cases, whereas Phokitis (1964) found 64 reports.

Among our 1405 patients with renal anomalies, only two (0.14 per cent) had a supernumerary kidney. The rarity of this anomaly and some specific features of its diagnosis warrant the presentation of the case record of one of them.

B., a 21-year-old male, was admitted to the clinic on September 2, 1958 complaining of rare dull pain in the lower abdomen which was referred to the pelvis. The pain was relieved when the patient lay on his back. He had been ill for four years. An elastic painless tumour, 6×10 cm in size, was palpated in the left iliac region. Blood and urine tests did not show any abnormalities. Chromocystoscopy revealed one orifice on each side of the urinary bladder; indigo carmine was discharged from them in the fourth minute. Excretory urography demonstrated the shadow of a pelvis on the left on a level with the fourth to fifth lumbar vertebrae with the calyces directed at the vertebrae; the shadow of a normal pelvis was seen in the

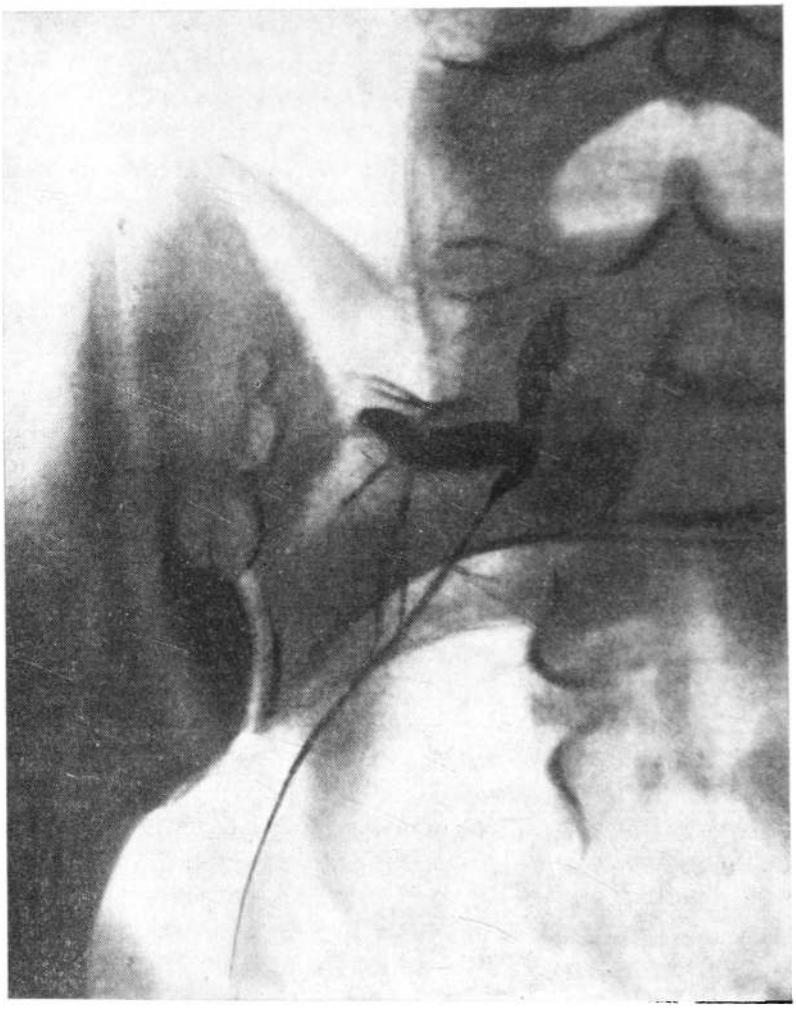


Fig. 112. Case B. A shadow of a rudimentary pelvis is seen on a level with the right sacroiliac articulation

right hypochondrium. There was also a shadow of an accessory rudimentary pelvis above the right sacroiliac articulation (Fig. 112).

Retrograde pyelography was undertaken. The shadow of a pelvis was seen on the left of the radiograph on a level with the fourth to fifth vertebrae; its calyces were directed laterally (Fig. 113). A short branch was seen arising from the right ureter above the right sacroiliac articulation and terminating in a rudimentary pelvis with two calyces. The details of the structure of the urinary tract of a supernumerary kidney shown by retrograde pyelography with high magnification are shown in Fig. 114.

The following diagnosis was made: iliac dystopia of a right supernumerary kidney with splitting of the right ureter; lumbar dystopia of the left kidney.

Treatment with spasmolytics and analgesics brought relief from the pain and the patient was discharged for out-patient care. Pain recurred now and again. He was readmitted to the clinic on July 12, 1970. As compared to the findings of the previous examination the function of both

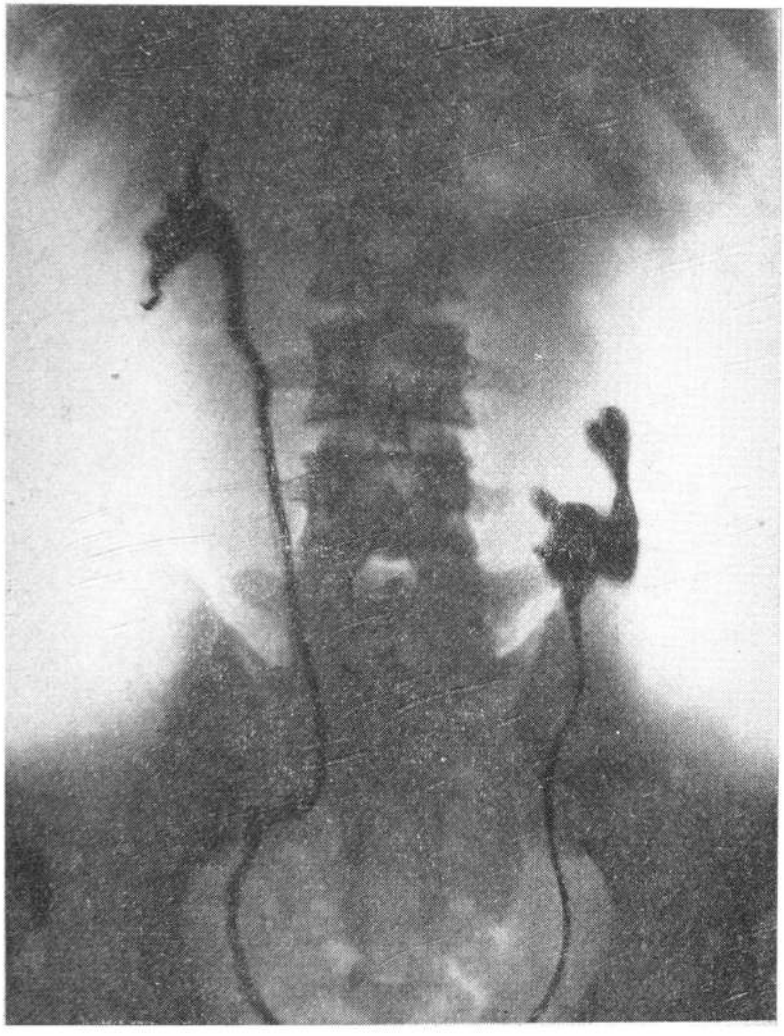


Fig. 113. The same patient as in Fig. 112. The right kidney is seen in the hypochondrium; lumbar dystopia of the left kidney. An accessory pelvis arises from the main ureter on the right at the level of the sacroiliac articulation. Retrograde pyelography

right kidneys was found to be diminished. The shadow of the ureter above the splitting was distended a little. Urine from the right ureteral orifice contained 20 to 30 leucocytes in field of vision. Culture of this urine produced growth of coli bacillus. The diagnosis of pyelonephritis of the supernumerary kidney and ureteral stricture at the site of splitting was made. Surgery was recommended but the patient did not agree to it.

Thus, in our case the rudimentary supernumerary kidney was found in the right iliac region. The right ureter of the supernumerary kidney separated into two branches, one terminating as the pelvis of the normal kidney in the right hypochondrium and the other, a short branch, being a rudimentary pelvis with

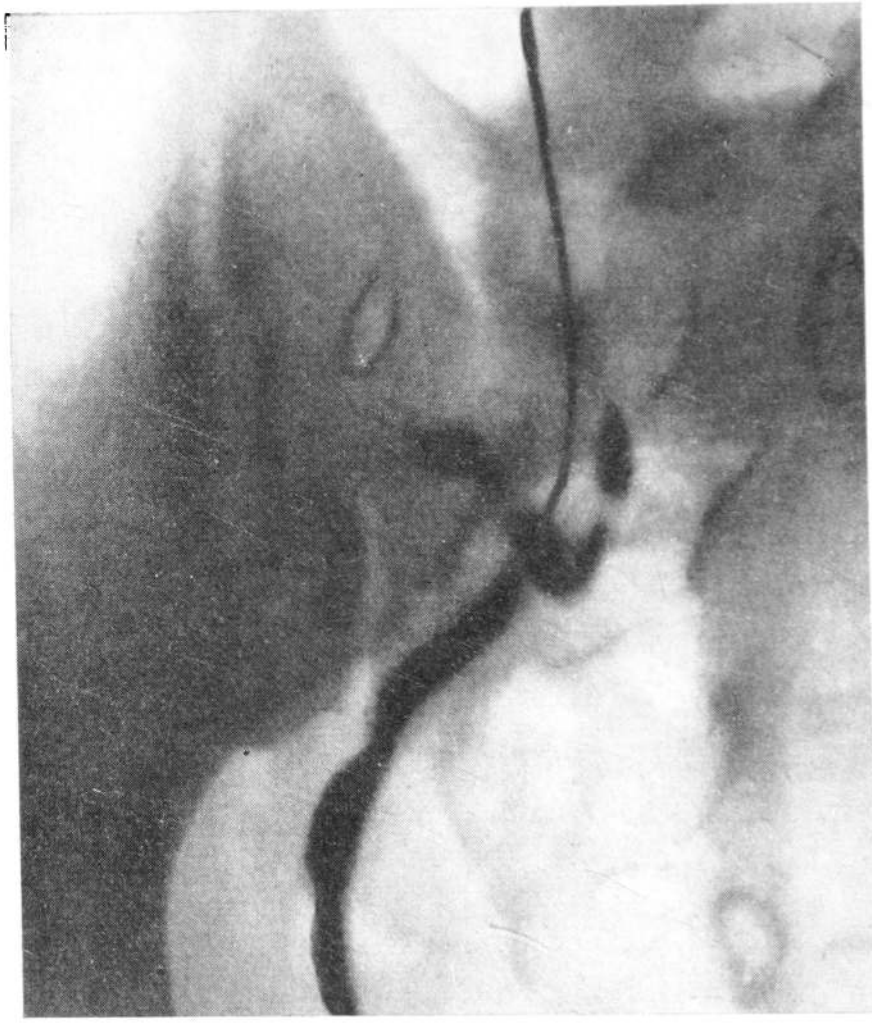


Fig. 114. The same patient. Retrograde pyelography. The shadow of the renal pelvis and two calyces of a supernumerary kidney is shown at high magnification

two calyces. Besides, the patient had a dystopic left kidney with the ureter terminating as an independent orifice in the usual place. Pyelonephritis of the supernumerary kidney had evidently developed.

According to the literature, a supernumerary kidney has been discovered from the age of nine months to 67 years, among males just as frequently as among females.

There may be more than three kidneys in this anomaly. Temelkoff (1957) described a confirmed case with four kidneys.

A 25-year-old man was admitted to a hospital for examination because of an abdominal tumour which caused pain of an indefinite character. An

egg-shaped, fixed and painful tumour of hard consistency was palpated close to the spine in the umbilical region. Blood and urine tests showed normal values. Laparotomy was carried out. A round elastic tumour, the size of a chicken egg and enclosed in a fibrous capsule, was removed from the retroperitoneal space. It was a kidney which had been situated on the abdominal aorta, from which its artery had arisen. Its ureter was joined to the ureter of the left kidney, which was normal in size and located in the lumbar region. Another tumour, the size of a nut, was found to the right of the aorta; its artery also branched off directly from the aorta. The ureter of this kidney terminated blindly. This kidney was also removed. Another, normal, kidney was found in the right hypochondrium. Histological examination of the resected kidneys showed normal renal structure but with elements of dystrophy.

This case is interesting not only because four kidney were found in one person, but also because the ureter of one of the supernumerary hypoplastic kidneys terminated blindly, owing to which the renal anomaly could not be recognized by the commonly used methods of pre-surgery X-ray examination.

Supernumerary kidneys differ greatly in size. In some cases they are small (the size of a bean or a chicken egg) and hypoplastic; in others they are almost normal in size. Most frequently they are small and function poorly. The main kidney on the side of the anomaly is often smaller than normal.

A supernumerary kidney is often pear-shaped or bean-shaped, although sometimes it has the appearance of a flattened mass. It is enclosed in a fibrous capsule. Frequently its surface is lobate, as in an embryonal kidney.

Histological examinations of supernumerary kidneys reveal different patterns. A normal histological structure with conspicuous cortical and medullary layers is the most common description in the literature. Small supernumerary kidneys, however, sometimes have a histological picture characteristic of hypoplastic kidneys.

The urinary tract of a supernumerary kidney resembles that of a duplicated kidney in structure. Each supernumerary kidney has its own ureter, which may be an independent structure opening into the urinary bladder by means of its own opening or a branch of the main ureter of the normal (main) kidney. Gimpelson (1945) noted that among summarized reports in half of the cases the ureter was split. Geisinger (1937) found this to be an even more frequent occurrence.

A supernumerary kidney often has an intrarenal pelvis; in exceptional cases it has two pelves and two ureters.

Descriptions can be found in the literature of a blindly terminating ureter of a supernumerary kidney and an ectopic ureteral orifice. According to Carlson (1950), this was the case in four of 51 summarized reports.

Each supernumerary kidney is separately supplied with blood from the aorta, whereas its veins drain directly into vena cava inferior. Quite often it has accessory vessels.

A supernumerary kidney is usually located below the main kidney, but is occasionally found above it. It is usually situated at the level of the lumbar vertebrae, less frequently in the iliac region or in the pelvis or in front of the promontorium. Carlson (1950) claims that a supernumerary kidney is found on the right just as frequently as on the left.

A supernumerary kidney is often found in conjunction with other renal anomalies. It may be dystopic, hypoplastic, duplicated or it may contain a solitary cyst. Anomalies of the contralateral kidney were noted in some cases. In the case described above a hypoplastic and dystopic right supernumerary kidney was combined with dystopia of the contralateral kidney.

According to the reports in the literature, a supernumerary kidney is discovered in many cases during examination for various diseases, e.g. pyelonephritis (Blagoveshchensky, 1931), nephritis (Parin, 1929), nephrolithiasis (Maksimovich, 1927), hydronephrosis (Sinovich, 1936). Summarized statistics published by these authors shows that these diseases develop very frequently in a supernumerary kidney. Other diseases are rarely encountered. Among these, malignant tumours should be distinguished; mixed teratoid tumours and carcinoma were found by Soroko (1930) and Skrychenko (1958).

This developmental renal anomaly has no characteristic symptoms. A supernumerary kidney of low localization may sometimes be palpated in the abdomen close to the vertebrae. This often creates difficulties in diagnosis and leads to unjustified surgical intervention. Other symptoms usually appear with the development of diseases and correspond to them, but in many cases the clinical picture is not typical of a renal disease. This is evidently linked to the low localization of the supernumerary kidney. Pain is often the first sign of the disease. The ailment usually takes a chronic course with periodical exacerbations. During exacerbation the disease of the supernumerary kidney has been mistaken for an attack of appendicitis, manifestations of ileus, diseases and tumours of the abdominal organs and female genitals.

A tentative diagnosis of this anomaly can be made when three or four separate kidneys not connected to one another are palpated. Sometimes the diagnosis was suspected when a urinary fistula formed on the side on which nephrectomy had been performed and the contralateral kidney was normal; such cases were encountered by Vashchinsky (1926) and Blagoveshchensky (1931). Because of injury suffered by the urinary tract the auth-

ors had to remove still another kidney from the same side, while the contralateral kidney was healthy. The presence of a supernumerary kidney should be suspected if there is an ectopic ureteral orifice and another two verified healthy kidneys which cannot be the cause of urinary incontinence.

The diagnosis of a supernumerary kidney is made on the basis of the findings of excretory urography or retrograde pyelography when two pelves are seen at a distance from one another and particularly when there are two separate kidney images. Pneumoretroperitoneum is advisable for obtaining a clearer image of the kidneys. These methods of examination make it possible to distinguish a supernumerary kidney from a duplicated kidney. Valuable information can be gained from renal angiography in cases in which it is difficult to make the diagnosis and particularly before undertaking surgery. Stefan (1956) produced on angiograms images of separate vessels, supplied to three kidneys, in the arteriographic phase and shadows of kidneys, not connected to each other, in the nephrographic phase.

The diagnosis is very difficult when there are only two orifices in the bladder and the supernumerary kidney functions poorly. It is no accident that, according to Carlson (1950), among 51 cases the correct diagnosis was made before operation in only four, during operation in 32, and during autopsy in 10; in five cases there were no indications when the diagnosis was made. Modern methods of examination allow the anomaly to be recognized if the surgeon bears in mind the possibility of its existence.

The question of treatment arises only in diseases of the supernumerary kidney. If there are no complaints, no treatment is required. Nephrectomy is the operation of choice in cases of disease. A small deficiently developed kidney has no functional importance and there is no need for an organ-preserving operation. Nephrectomy is also advisable when the deficiently developed supernumerary kidney is found by chance during operation, but the surgeon must be sure that the patient has normal kidneys.

In conclusion to this chapter, we would like to emphasize that renal anomalies in number are made up of malformations differing in embryogenesis, functional importance and morphological features; these are agenesis and aplasia of kidneys, hypoplastic, duplicated, and supernumerary kidneys.

The clinical importance of this group of developmental anomalies is considerable. It is linked first of all with the fact that such kidneys are often encountered in clinical practice. This particularly applies to duplicated kidneys, which account for 3.4 per cent of all patients treated at urological clinics for diseases

of the kidney and upper urinary tract and for 45.6 per cent of all encountered renal anomalies. Renal agenesis and aplasia account for 0.18 and 1.28 per cent and hypoplasia for 0.58 and 2.7 per cent, respectively. Supernumerary kidneys are a very rare occurrence.

These developmental anomalies of the kidneys are sometimes the cause of tragic mistakes (removal of a solitary kidney in aplasia or the only adequately functioning kidney in hypoplasia) and unjustified operations due to diagnostic errors. They may be the cause of urinary fistulas which form when the upper urinary tract of an unrecognized duplicated or supernumerary kidney is injured during operation.

These anomalies are also important clinically because they often cause pathological phenomena. Pain, haematuria, and arterial hypertension may be associated with disturbed urodynamics in the upper urinary tract in anomalies in the number of kidneys. Dyskinesia of the upper urinary tract is revealed particularly clearly by urokymography and urotelescopy. Examination shows that in 8 per cent of people with duplicated kidneys and no disease urodynamics of the upper urinary tract is impaired and causes urinary stasis. The causes of dyskinesia lie in the anomalous anatomical structure of these kidneys. The same causes evidently underlie the predisposition of such kidneys to diseases linked with urinary stasis in the pelves, such as hydronephrosis, pyelonephritis, and nephrolithiasis. These account for 84.5 per cent of diseases of duplicated kidneys. They are often encountered in a solitary, hypoplastic and supernumerary kidneys. The disease in such cases usually follows a chronic course.

Anomalies in the number of kidneys create diagnostic difficulties because not only the disease of the kidney has to be diagnosed but also its developmental anomaly, and their clinical and X-ray signs have to be distinguished. In addition, the clinical picture of the disease in such cases is often atypical because of the abnormal position of the anomalous organ and the combination of the symptoms of the anomaly with those of the disease. It is very important to search for a renal anomaly in each case, and particularly when developmental anomalies of the female genitalia or other organs are discovered. Such combined anomalies are encountered particularly often in cases of renal agenesis and aplasia.

The diagnosis of an anomaly in the number of kidneys can be made by means of cystoscopy, excretory urography, retrograde pyelography and pneumoretroperitoneum in almost all cases. Renal angiography is sometimes needed if it is difficult to make a diagnosis. The functional condition of the kidneys is judged according to the findings of chromocystoscopy, renog-

raphy, kidney scanning and excretory urography, whereas the function of the ureters is studied by means of urokymography and cineurography.

* * *

The clinical significance of these renal anomalies is also seen in the specific features of the management of their diseases. Some of the anomalies themselves require surgical treatment. Surgery should be considered when a hypoplastic kidney causes pain and arterial hypertension. Surgery is necessary in cases of blind supernumerary ureters, ectopic ureteral orifice, and ureterocele; the last condition is encountered quite frequently in cases of duplicated kidneys (in 8.9 per cent of all cases). In marked dyskinesia of the upper urinary tracts of duplicated kidneys an early operation performed before a disease develops or renal function diminishes yields better results. Surgery is also often applied to treat diseases of this group of anomalous kidneys, particularly hydronephrosis, nephrolithiasis, and pyelonephritis.

Operations in which the organ is preserved as much as possible are usually performed on these anomalous kidneys. Nephrectomy is expedient in various diseases of a deficiently functioning hypoplastic kidney, of both segments of a duplicated kidney or in diseases of supernumerary kidneys with disturbance of their function, as well as in tumours of an anomalous kidney. In all other cases it is preferable to preserve the kidney or part of it.

Heminephrectomy is the operation of choice in various diseases of a duplicated kidney (with the exception of tumours) with marked functional and morphological changes in one of its segments, and in blind termination of the distal segment of the ureter of a duplicated or supernumerary kidney with impaired function.

The whole duplicated or supernumerary kidney may be preserved by means of pyeloureterostomy or uretero-ureterostomy. These reconstructive operations are indicated in cases of disturbed urodynamics in the upper urinary tract, in hydronephrosis, nephrolithiasis or pyelonephritis, in ectopic ureteral orifice, in vesicoureteral reflux into one of the two ureters; in reflux into the common ureter the anastomosis is combined with an antireflux operation. Such operations should be undertaken only on condition that the function of the kidney is preserved. We recommend removal of the whole non-functioning ureter during the operation so as to prevent disturbance of the urodynamics of the remaining ureter. This is why the best results are

obtained when the anastomosis is formed at a high level. We cannot speak in favour of an operation to form anastomosis between the ureters of a duplicated kindey and the bladder since it does not correct the urodynamic disorders and does not remove the causes of urinary stasis in the upper urinary tract. We likewise do not find it expedient to ligate one of the ureters in ectopia of the orifice. This operation leaves the focus of infection in the anomalous kidneys.

CHAPTER VI

Ureteral Anomalies

It is impossible to suggest an exact classification of ureteral anomalies today which would accord with current theoretical knowledge.

Four groups of developmental anomalies of the ureters can be distinguished.

I. Anomalies in the number of ureters, including agenesis and duplication and multiple ureters.

II. Anomalies of the proximal or distal ureteral segment, including ectopia of the orifice, ureterocele, and blind termination of ureters.

III. Anomalies of position, including retrocaval ureter and herniation of the ureter.

IV. Anomalies in the calibre, shape, and structure of the ureters, including aplasia and hypoplasia, congenital strictures, valves, diverticula of the ureter, spiral ureter, congenital dilatation of the ureters (primary megaloureter, primary atonia and neuromuscular dysplasia of the ureter).

Pytel and Chebanyuk (1969) distinguish the following groups of ureteral malformations: anomalies in number, anomalies of initial and terminal segments, and anomalies in shape and structure. Retrocaval ureter, herniated ureter and congenital ureteral dilatation due to neuro-muscular dysplasia do not fit into this classification.

Ureteral agenesis is due to an undeveloped ureteral bud. This developmental anomaly usually occurs in conjunction with aplasia or agenesis of the kidney and therefore has no independent significance.

Duplication of the ureter is a relatively frequent anomaly. Two variants are encountered—complete duplication, in which each ureter has its own orifice and pelvis; and incomplete duplication, when the ureter divides into two branches above the orifice and each branch has its own pelvis. The splitting occurs most frequently in the upper third of the ureter, less frequently in the middle third, and still less frequently in the lower third. Duplication of the ureter can be unilateral or bilateral. This type of anomaly is mostly found in females. It develops from two ureteral buds of the nephrogenous blastema or due to the division of one ureteral bud. Cases have been described in the literature of two ureters joining at the pelvis to form a single ureter.

In cases of complete duplication two orifices are found in the urinary bladder, the upper one corresponding to the lower pelvis and the lower orifice corresponding to the upper pelvis (Weigert-Meyer's law). There are exceptions to this rule, though they are rare (Lund, 1949).

Duplicated ureters often cross one another, sometimes more than once. The unfavourable structure of duplicated ureters is thus conducive to the development of urodynamic disorders. These lead to a certain extent to functional obstruction of the upper urinary tract manifested in increased intrapelvic pressure and pain.

Depending on their degree and duration these disorders cause retention diseases such as hydronephrosis, pyelonephritis, and nephrolithiasis.

The anatomical preconditions for the development of functional disorders and diseases are the close contact of both ureters situated in a common seat, defects in the ureteral orifice and the site of ureteral splitting, scars, valves, strictures, fixed ureteral kink, compression of the ureters by accessory vessels, and various developmental malformations of the terminal part of the ureter. Anatomical and functional changes in the vesicoureteral segment are among the important factors promoting the development of vesicoureteral reflux (VUR) in cases of duplication of the urinary tract. The congenital character of this anomaly is confirmed by its frequent occurrence in conjunction with duplication of the urinary tract and deficient termination of the ureter. This deficiency is encountered only in the intravesical segment of the ureter, whereas the proximal parts of both ureters are normal.

The anatomical preconditions for vesicoureteral reflux into the ureter of a duplicated kidney are lateral displacement of the orifice; the frequently encountered ureterocele of the accessory ureter causing abnormal interrelationships in the region of the

trigone of the bladder; and a shortened intravesical segment of the ureter of the lower part of the duplicated kidney.

This explanation for the appearance of pain and the subsequent development of disease is confirmed by our observations using urotelescopy and cineurography.

In duplication of the urinary tract the ureters contract in different rhythms, asynchronously. During contraction of the pelvis a small amount of the contrast medium is pushed into the ureter while the main mass returns into the calyces and distends them. In ureteral splitting, interureteral reflux is often seen and pendulum-like movement of the fluid from one ureter into the other. This reflux was encountered in several variations in our cases. In some the radiocontrast medium moved from the lower pelvis into the upper pelvis through the site of the splitting and partly into the common ureter, and then after contraction of the upper pelvis and its ureter it flowed into the common ureter. In other, rarer, cases some of the radiocontrast medium flowed from the upper pelvis through the site of the splitting into the common ureter, but the bulk of it entered the lower pelvis and, on contraction of the latter, was pushed into the common ureter. There were also patients in whom the medium moved pendulum-like from one pelvis into the other and back again, and with each such movement a small amount of it was pushed into the common ureter.

The diagnosis of duplicated urinary tracts and their diseases is based on the findings of cystoscopy, during which accessory ureteral orifices are seen, excretory urography, demonstrating duplicated urinary tracts and changes in them; and urotelescopy and cineurography, by means of which the dynamics of the urinary tract is disclosed. All other examination methods are of auxiliary value.

The diagnosis of VUR in duplication of the urinary tract, just as in all other cases, is based on the results of cystoscopy, namely the discovery of the lateral position of the orifice, a gaping orifice, and a shortened intravesical ureteral segment. Cystourethrography helps in revealing passive and active reflux and the extent of the vesico-ureteral-renal reflux (Dzhavad-zade et al., 1977). In addition to these methods of examination infusion excretory urography is also always undertaken. If the bladder is atonic, trabecular in structure and contains residual urine, it is necessary to make sure that there is no intravesical obstruction and no neurogenous bladder; this will help in planning the sequence and character of surgical manipulations in VUR in general and in duplication of the upper urinary tract in particular.

The management of diseases in duplication of the upper urinary

tract is discussed in Chapter V in the part dealing with diseases of a duplicated kidney. Here we focus the reader's attention on the treatment of VUR in duplication of the upper urinary tract.

The indications for non-operative or surgical treatment, as well as the choice of the type of the operation, are determined to a great measure by the character and extent of the disturbed function of the ureteral antireflux mechanism and by the factors conducive to the development of VUR in each case. Treatment consists in removing the factors contributing to the development of VUR, infection of the urinary tract, and regurgitation of urine. In the absence of functional disorders in the upper urinary tract an attempt is made to correct VUR by means of non-operative measures. Surgery should not be delayed if the orifice gapes and if the intravesical segment of the ureter is absent or shortened and passive vesico-renal reflux occurs at a high level. The young age of a child is not a contraindication for surgery. Surgery is unquestionably indicated when coarse irreparable morphological changes are found in the upper urinary tract.

The following circumstances are taken into account in choosing the surgical method: the functional condition of the involved part of the kidney; the presence of reflux in the neighbouring or contralateral ureter; the type of duplication; the condition of the terminal ureteral segment; the presence and level of activity of inflammatory processes. It is advisable to undertake the operation during a remission of inflammation.

Lack of function in the affected part of the duplicated kidney is an indication for heminephro-ureterectomy with total removal of the refluxing ureter. Nephro-ureterectomy has to be resorted to when there are considerable changes in both segments of the kidney, in which case the condition of the contralateral kidney must be taken into account. VUR of the contralateral ureter with bilateral dilatation of the upper urinary tract calls for an organ-preserving operation carried out in stages. The first operation is performed on the better functioning kidney.

The expediency of antireflux operations on a refluxing ureter in duplication of the urinary tract is still debatable. Ledbetter-Politano ureterocystoneostomy in duplication of the upper urinary tract is often not radical enough because it does not correct the urodynamic disorders in the anomalous ureters.

These disorders of urodynamics may contribute to the maintenance of infection in one of the renal segments even if there is no VUR. To provide for good drainage of urine from the upper urinary tract in three cases we performed ureteroneopyelostomy after we had removed an accessory refluxing ureter; the operations were successful.

In duplication of the urinary tract transvesical surgery harbours considerable risk because the orifice of the second ureter, found close to that of the refluxing ureter, may be damaged during the intervention. For this reason we consider this operation very dangerous in this anomaly. If it cannot be avoided, however, it is advisable to catheterize both ureters for better orientation. 'Singleblock' antireflux surgery for VUR in both ureters, which is recommended by some authors, is simple in technique but the disorders of urodynamics associated with the unfavourable anatomical and functional condition of the anomalous ureters (their asynchronous contractions) cause disturbances in the passage of urine.

The presence of isolated segments of the duplicated upper urinary tracts makes it possible to replace the refluxing ureter with the healthy one or to remove one of the two refluxing ureters. Ureteroneopyelostomy or interureteral anastomosis carried out at a high level provides for the normal flow of urine by way of the one remaining ureter. In cases of VUR into both ureters, we divided one of the refluxing ureters and performed ureteroureterostomy or ureteroneopyelostomy and simultaneously performed an antireflux operation on the other remaining ureter by means of a modified Ledbetter-Politano technique. Such operations have good remote results, as was the case in nine of our patients. In one, pyeloureterostomy was a component of reconstructive surgery on a horseshoe kidney. The isthmus of the kidney was resected, one of the ureters on the left into which the VUR occurred, was removed, and pyeloureterostomy was carried out. The terminal part of the refluxing ureter must be removed without fail in these operations.

It is our experience that pyelonephritis of the affected segment of a duplicated kidney is not a contraindication for such organ-preserving operations. Surgical correction of urodynamic disorders is only a component of pyelonephritis management. Later, after the operation, such patients require continuous long-term antibacterial therapy according to the schedule commonly accepted in the treatment of this disease.

Multiple ureters are relatively rare occurrence. In our experience they accounted for approximately 1 per cent of cases of duplication of ureters. Multiple ureters can be found on one or both sides. Campbell- Begg (1953) had a female patient with four ureters on the left and two ureters on the right side.

Anomalies of the proximal and distal parts of multiple ureters include ectopia of the orifice, ureterocele, and blind termination of the proximal or distal segment. The clinical picture, diagnosis and treatment of diseases in these cases do not differ from those in duplication of the upper urinary tract.

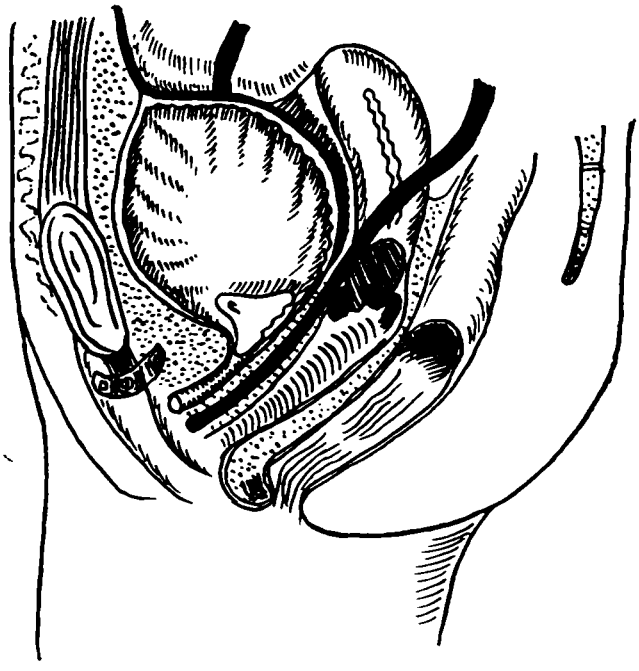
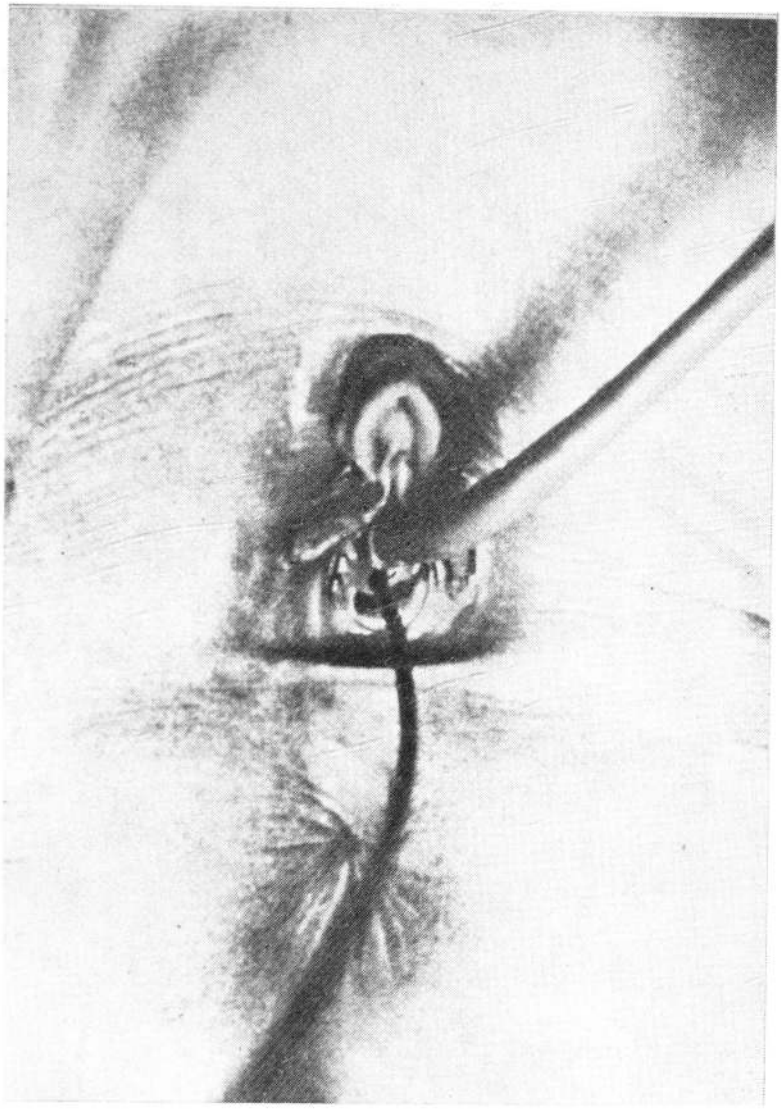


Fig. 115. The external genitals of a girl. A rubber catheter is introduced into the urethra and a ureteral catheter into the ectopic orifice. A diagram of the position of the ectopic ureteral orifice is shown on the left

Ectopia of the ureteral orifice. In this anomaly of the distal part of the ureter, its orifice is found below the trigone of the bladder. Edinyi (1953) distinguishes two groups of ectopia of the ureteral orifices from the standpoint of embryogenesis. The first group includes ectopia of the orifice into organs arising from the urogenital sinus, e.g. cervicovesical, urethral, and paraurethral (vestibular) ectopia. The second group embraces ectopia into organs arising from Mueller's ducts and intestinal tube, i.e. into the vagina, uterus, and intestine. Two groups are also distinguished from the standpoint of their clinical manifestations: suprasphincteral and infrasphincteral ectopia. In suprasphincteral ectopia the ureteral orifices are often found in the neck of the bladder and the urine is therefore retained in the urinary bladder. In infrasphincteral ectopia involuntary passage of urine, enuresis ureterica, is noted. The disease is mostly encountered among females, in whom there are both urinary incontinence and voluntary passage of urine. All other symptoms are usually associated with dilatation of the proximally located urinary tract and infection.

An ectopic ureteral orifice may be detected by inspection,



cystourethroscopy, urethrography, excretory urography, and retrograde pyelography. It is very difficult to diagnose this anomaly in males because the ectopic orifice may be located in the posterior urethra, vas deferens, seminal vesicles, epididymis or rectum. In such cases pain in the corresponding region, dysuria, pyuria, and dull lumbar pain are observed. When ectopia of the orifice into these organs is suspected and there is a cyst-like structure, a puncture is made, fluid is aspirated for examination, and a radiocontrast medium is then injected. Cervical and posterourethral ectopia of the orifice is found by means of cystourethroscopy, with the administration of fluid into the bladder and with elimination of fluid from the bladder, and obligatory intra-

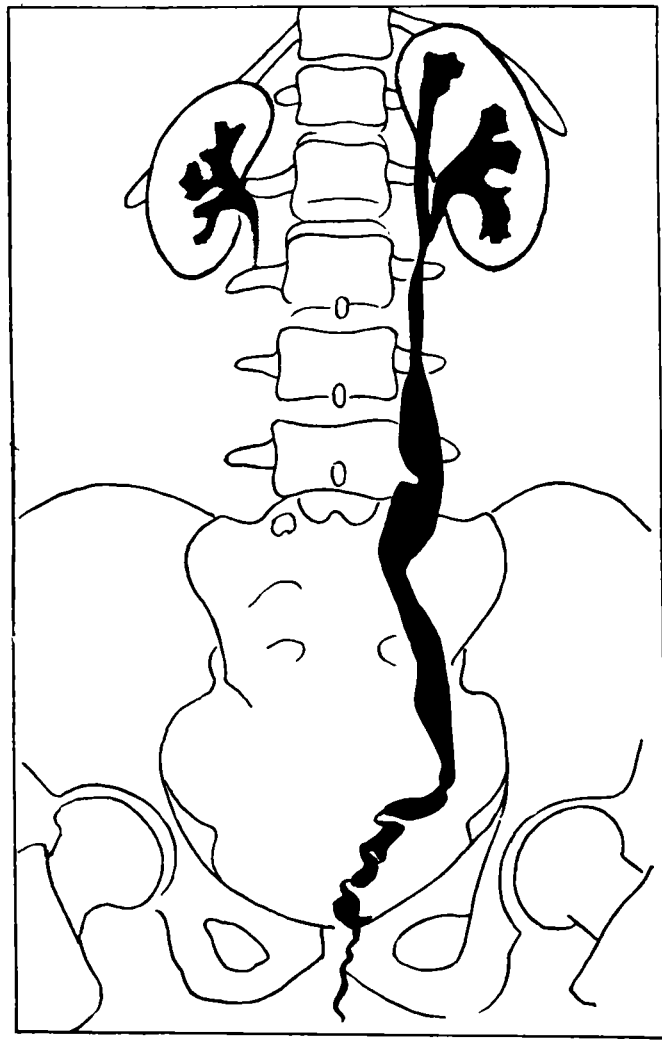
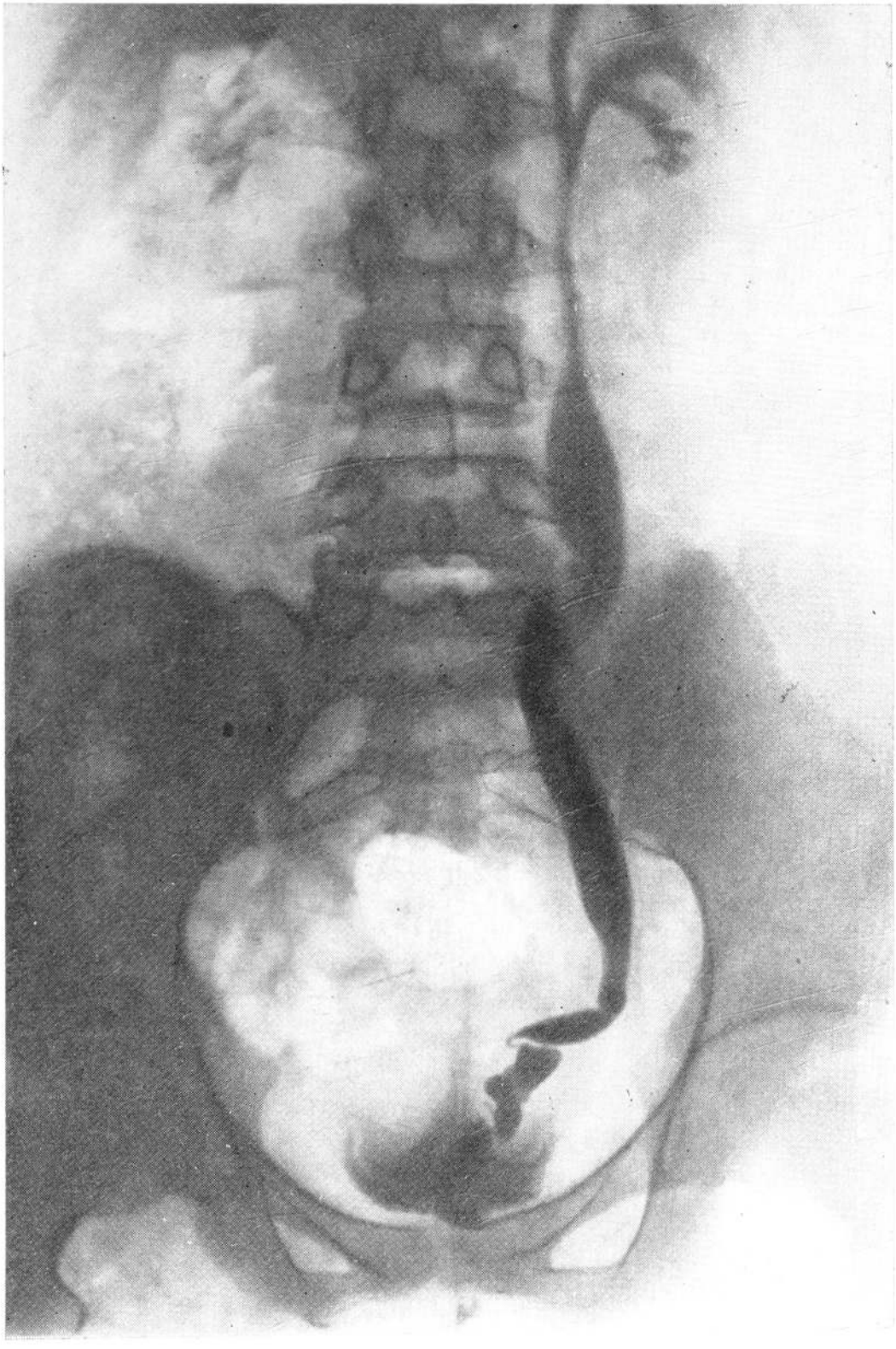


Fig. 116. Excretory urogram with ascending ureteropyelogram on the left. Complete duplication of the upper urinary tract on the left with ectopia of the ureter of the upper segment

venous infusion of indigo carmine. In this method the neck of the bladder and the posterior urethra are dilated by the fluid stream (Ismailov).

In a female, urinary incontinence may not occur when she is lying down. The diagnosis in females can be also made late, because the symptoms may be mistaken for those of enuresis, vaginitis, etc. The diagnosis is first made from the results of the indigo carmine test and then from the findings of excretory and retrograde pyelography (Fig. 115).

In ectopia of the ureteral orifice the proximal parts of the ureter are dilated; Pytel and Chebanyuk (1969) attribute this



dilatation to deficient development of the neuro-muscular apparatus. It should be noted that the dilatation is usually caused by strangulation of the ureter by the surrounding tissues and by the periodical filling and emptying of the upper urinary tract. This is confirmed by our cases (Fig. 116).

Treatment depends to a great extent on the functional and morphological changes in the kidney and ureter and on the number of ureters on the relevant side. In ectopia of the orifice of one of the duplicated ureters, with mild ureteral dilatation and the corresponding renal segment intact, pyelo-pyelostomy, pyeloureterostomy or uretero-ureterostomy are advisable; these operations produce better results than ureterocystoneostomy. We performed them on all of our patients with good remote results. We prefer side-to-side uretero-ureterostomy to the end-to-side method because the technique is simpler and the resulting anatomical relationships better (Fig. 117). Heminephro-ureterectomy is indicated in cases of considerably diminished function of the renal segment with the atonic ureter. The antireflux variant of ureterocystoneostomy is undertaken if the urinary tract is not duplicated.

The indications for nephro-ureterectomy do not differ from those generally accepted.

Blind termination of the ureter in the proximal or distal part is very rare. Blind termination of the proximal segment in ureteral duplication is sometimes accepted as and called ureteral diverticulum. The orifices of such an ureter can be found in the usual place. Voino-Yasenetsky (1962) believes that a similar picture can be encountered in strictures of the ureter but in such cases the flow of urine in the ureter is not obstructed. A blindly terminating ureter sometimes resembles a fibrous strand and the kidney in such cases is aplastic or markedly hypoplastic. The diagnosis and clinical picture of the anomaly are discussed in Chapter V. Figs. 96, 97, and 98 show a typical picture of blindly terminating ureters. Surgery for blindly terminating ureters comprises ureterectomy or heminephro-ureterectomy.

Ureterocele. In ureterocele the ureteral orifice is stenosed and a cyst-like dilatation of the intravesical ureteral segment develops. This dilated part is covered on the outside with the mucous membrane of the bladder and lined inside with the ureteral mucosa. Between these two mucous layers muscular and connective-tissue elements are arranged diffusely because they are drastically stretched.

In the initial stage of the disease, when the ureterocele is small, the upper urinary tract is not dilated. With the development and enlargement of ureterocele the dilatation spreads to the distal parts of the ureter and, finally to the pyelo-calyceal system,

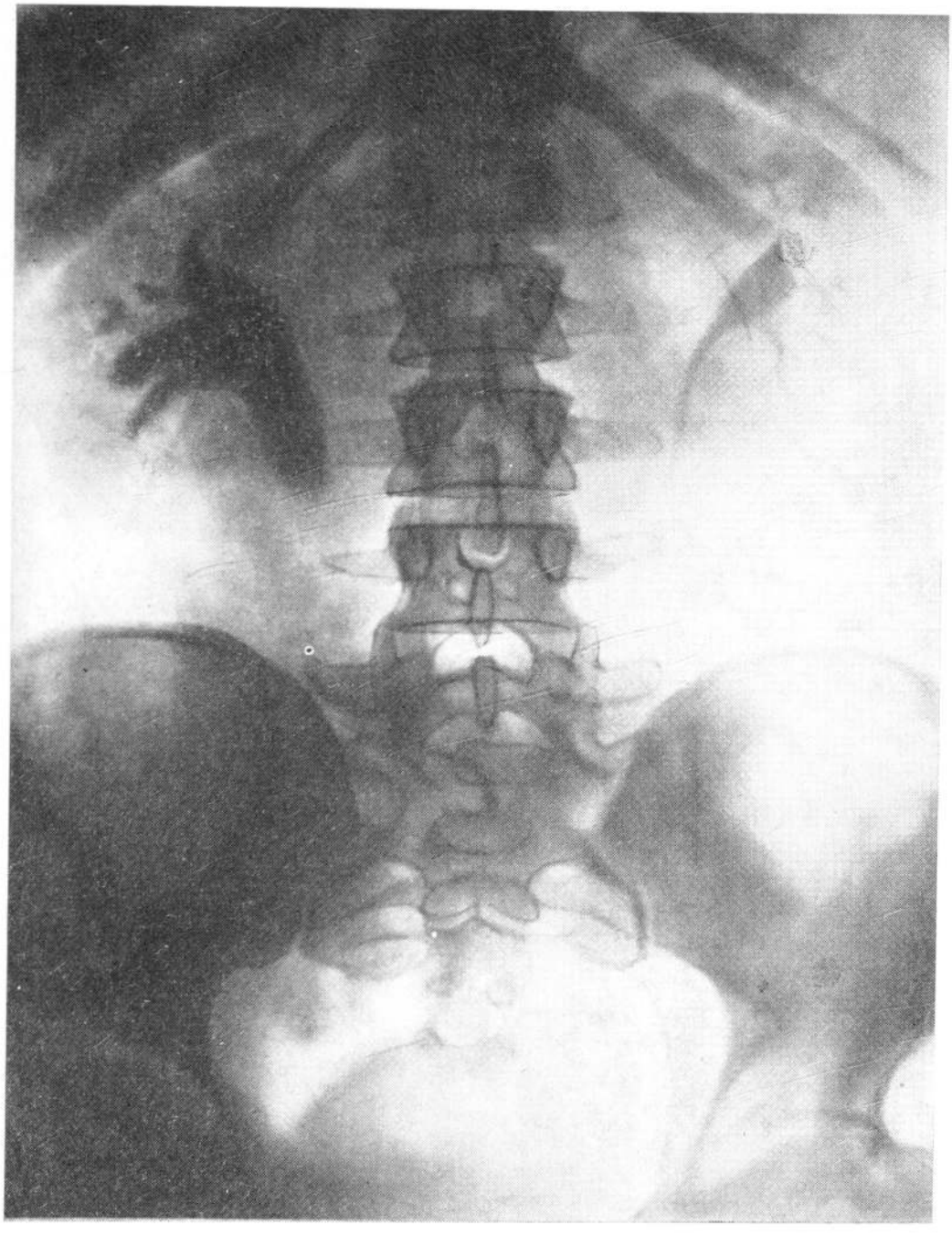


Fig. 117. Excretory urogram of the same patient as in Fig. 116 made after side-to-side uretero-ureterostomy with resection of the peripheral segment of the ectopic ureter

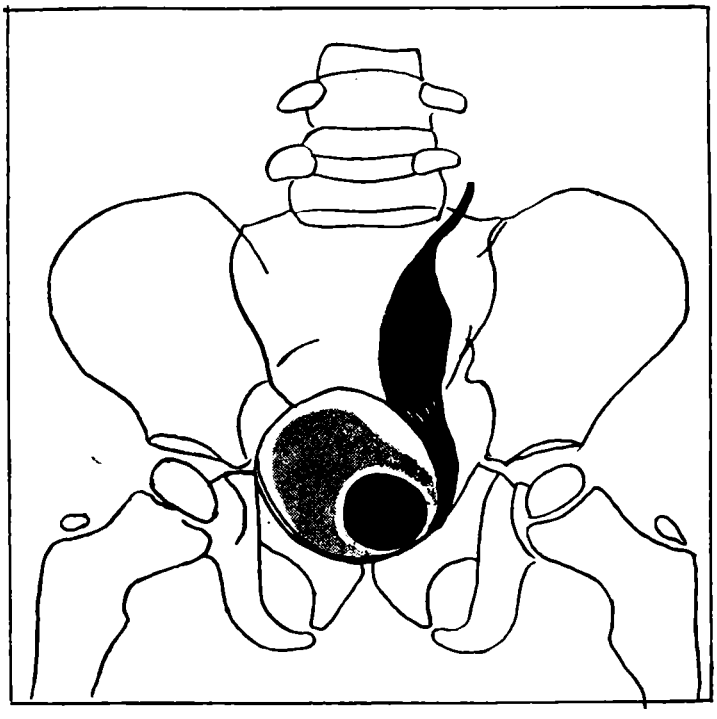
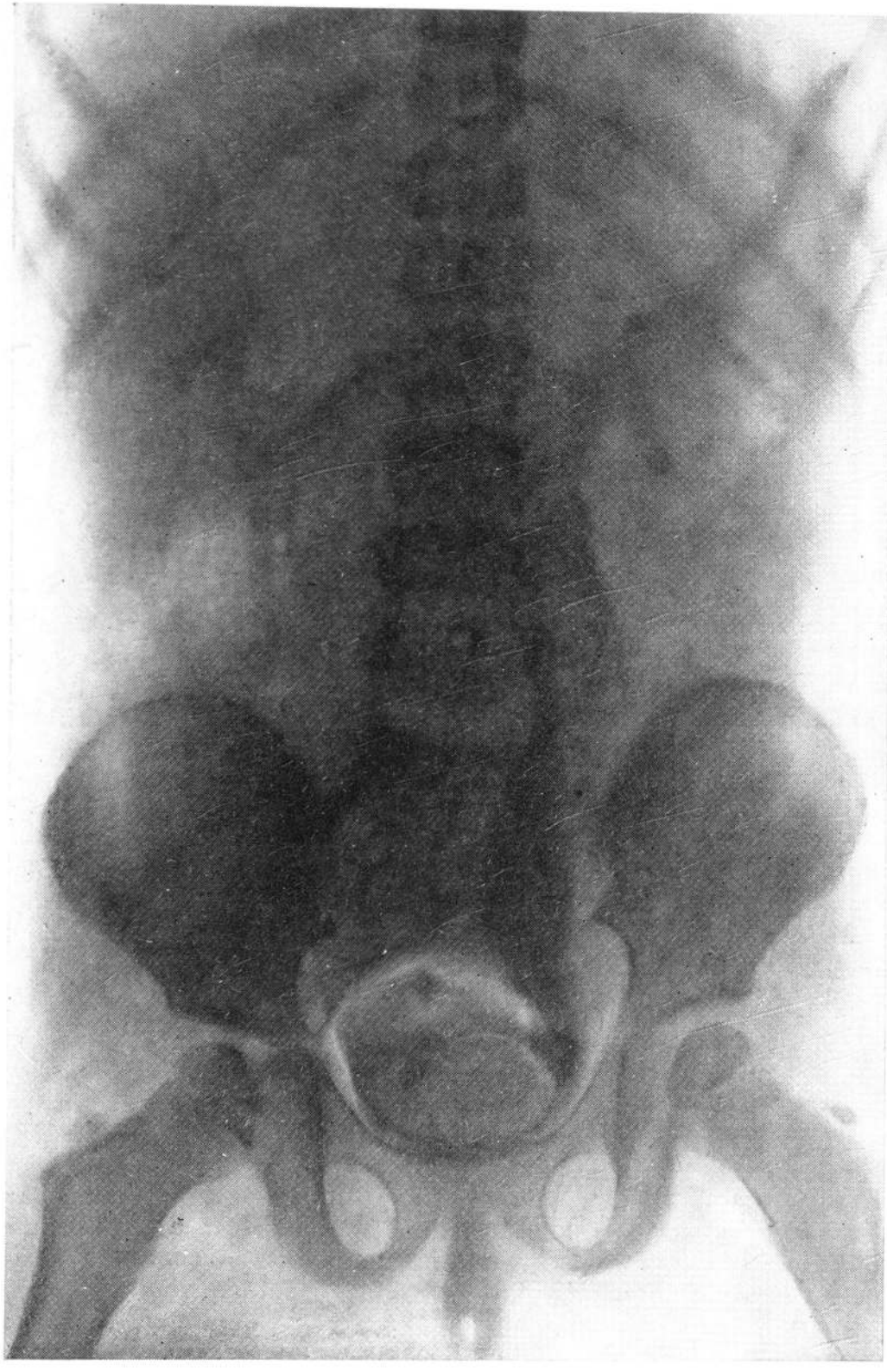


Fig. 118. Excretory urogram with descending cystogram (a large filling defect is seen in the bladder). A diagram of descending cystogram is shown on the left

with subsequent atrophy of the renal parenchyma, the picture is that of hydro-ureteronephrosis with all its consequences.

The size of ureterocele is determined by the length of the intravesical ureteral segment and the degree of stenosis of its orifice, as well as by concomitant infection. The intravesical part of the ureter is distended with the urine and may floatate. In the latter case ureterocele closes the internal urethral orifice, i.e. the neck of the bladder, during urination and is one of the causes of obstructive diseases of the infravesical region. We had two girl patients with prolapse of ureterocele through the urethra; similar cases are often reported in the literature. The disease may be uni- or bilateral and is encountered more often among girls. It is revealed in 4 per cent of children examined for pyuria (Campbell, 1951). It should be noted that in duplication of the upper urinary tract ureterocele is usually found in the accessory ureter.

The condition of the upper urinary tract and kidney is very important in determining the clinical course taken by ureterocele. That is why Usou's classification (1961), compiled with due account for the degree of involvement of the proximally situated parts of the urinary tract, deserves attention. It distinguishes (1)



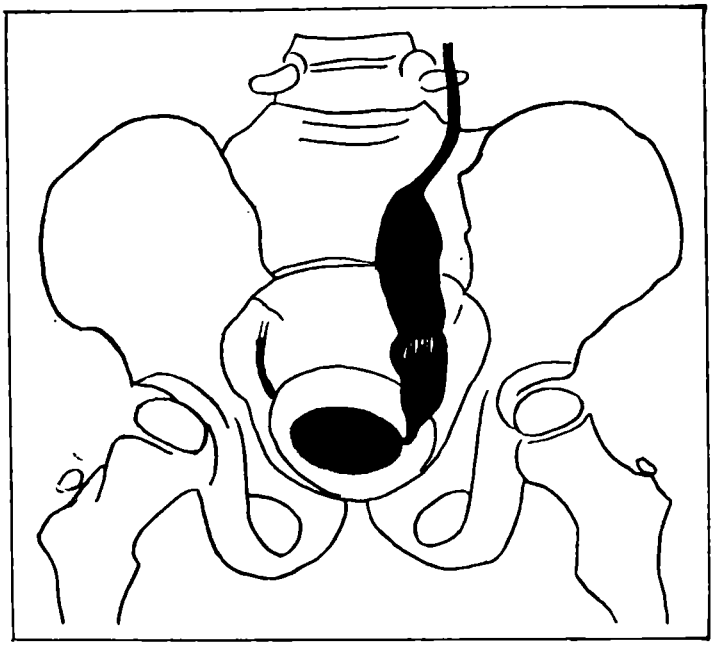


Fig. 119. Excretory urogram after evacuation of the bladder. The contrast medium fills the ureterocele completely. A diagram of the same case is shown on the left ▶

small ureterocele with minimum involvement of the upper urinary tract; (2) large ureterocele with involvement of the ureter and kidney; (3) large ureterocele with involvement of the ureter and kidney, causing micturition disorders because it penetrates the neck of the bladder. The symptoms of ureterocele are associated with stasis of urine in the upper urinary tract and infection and, in the third group, with retention of urine in the bladder. Dysuria, pyuria, proteinuria, and frequent elevation of body temperature are characteristic manifestations. Ureterocele follows a particularly severe course in infants (one to four months of age) with dyspepsia and marked toxicosis (Ostropolskaya and Dolgatov, 1975).

The diagnosis of ureterocele presents no difficulties and is based on the findings of cystoscopy, cystography and excretory urography. Ostropolskaya and Dolgatov (1975) recommend excretory urography and claim that a spherical filling defect is demonstrated in the bladder immediately the contrast medium flows into it. One cannot agree with this opinion, since renal function is reduced to some degree in cases of ureterocele and, besides, ureterocele is often encountered in duplication of the upper urinary tract in the accessory ureter, and, consequently, cannot be filled with the contrast medium immediately after intravenous infusion.

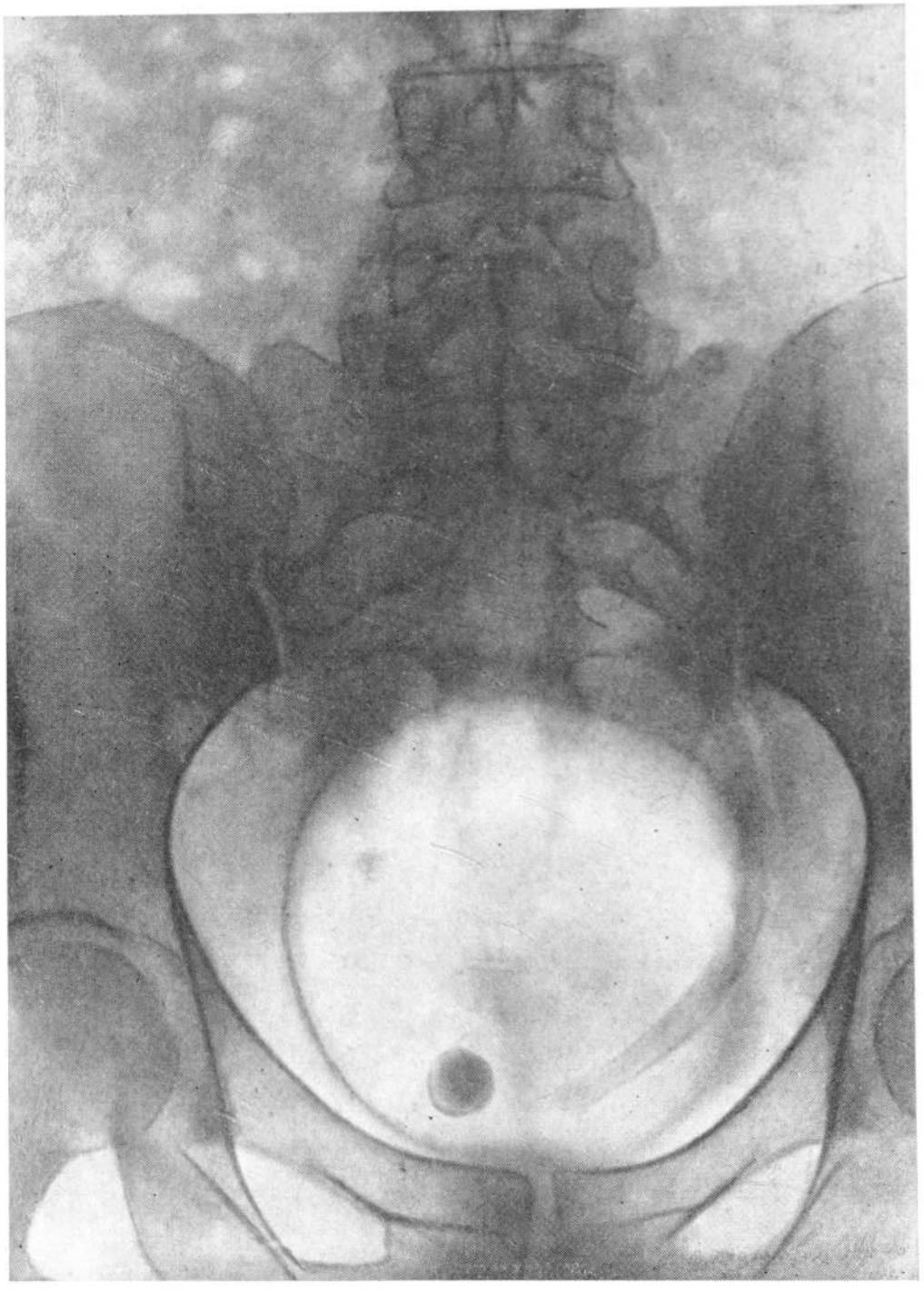


Fig. 120. Descending cystogram. Bilateral ureterocele. Stones in the dilated intravesical ureteral segment are seen on the right

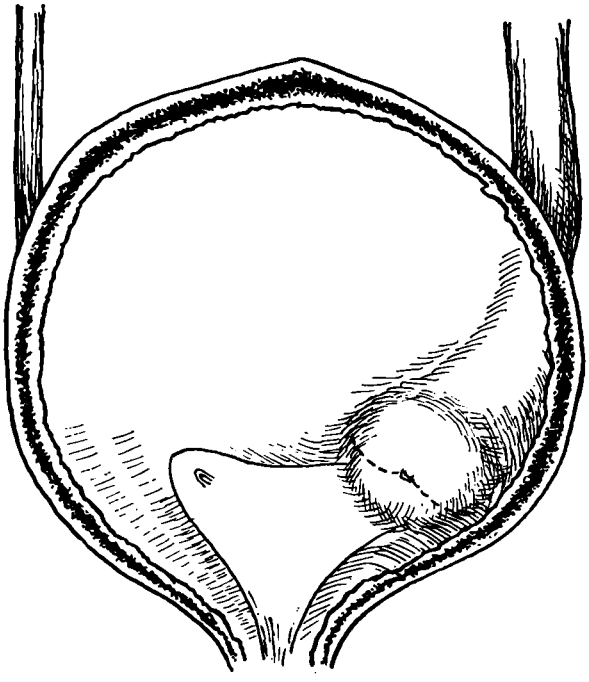


Fig. 121. Diagram of ureterocele. The dashed line shows the direction of the incision made through the orifice

When renal function is reduced, large ureterocele is demonstrated on a descending cystogram as a filling defect (Fig. 118). To demonstrate ureterocele on a delayed descending cystogram, the urinary bladder is evacuated and oxygen is administered. The shadow of ureterocele filled with the contrast medium is seen against the oxygen background (Figs. 119 and 120). There is no doubt that excretory urography must be undertaken in order to determine the condition of the upper urinary tract, but cystoscopy is the main diagnostic method. Its diagnostic value is less only in cases of large ureterocele occupying the whole cavity of the bladder. It should be noted that sometimes the shadow of ureterocele may not be shown by ascending cystography, in which case a small amount of low-concentration contrast medium is introduced so as not to compress the ureterocele.

Ureterocele is managed by surgery and the following operations are suggested for this purpose: (1) transureteral division or coagulation of the ureterocele opening; (2) transvesical division of ureterocele; (3) transvesical excision or removal of ureterocele with antireflux protection; (4) nephro-ureterectomy; (5) hemineph-ro-ureterectomy; (6) uretero-ureterostomy with removal of ureterocele; (7) ureteroneopyelostomy with removal of ureterocele.

The choice of the operation is determined by the size of the ureterocele, the extent of the dilatation of the upper urinary tract, the number of upper urinary tracts, the bi- or unilateral character of disease, the possibility of vesico-renal reflux, and the condition of the lower urinary tract.

Transurethral or transvesical division of ureterocele is indicated if it is small and the urinary tract and kidney function satisfactorily. In such cases the ureteral orifice is cut not upward, as it is suggested by many authors, but medially or laterally, as is done by Ismailov (Fig. 121). After transvesical excision or removal it is necessary to perform an antireflux operation such as ureterocystoneostomy or Bischoff's transvesical operation. An ectopic ureterocele located at a very low level deserves particular attention because regurgitation of urine may occur when it is divided or excised, even despite its small size. Such patients are examined again for VUR.

Ureterocele causing complete destruction of the intravesical parts of the ureter, even when there is only mild dilatation of the upper urinary tract, must not be excised or divided, because regurgitation of urine is inevitable in such cases. This is why such ureterocele requires antireflux protection.

Nephrectomy in cases of ureterocele of a solitary ureter (on the corresponding side) or heminephro-ureterectomy in cases of ureterocele of one ureter of a duplicated kidney is indicated for patients with marked changes in the upper urinary tract and kidney. Heminephro-ureterectomy in duplication of the ureter with atonia of the urinary tract and pyelonephritis protects the other half of the kidney from the development of inflammation.

High-level uretero-ureterostomy or ureteroneopyelostomy is indicated when renal function on the corresponding side is satisfactory. These operations require that the ureter and its cystic dilatation are removed. We pay special attention to this manipulation because the main ureter may be damaged during removal of the stump of the accessory ureter. This is particularly so when the lower segments of both ureters cross. It is our experience that one cannot count on spontaneous collapse of ureterocele in these operations, which is why we think it advisable to excise the ureterocele and the adjoining thinned wall of the bladder so as to prevent eventual sacculation there. This sacculation, like ureterocele or diverticulum of the bladder, may be an obstacle for the flow of urine from the orifice of the remaining ureter found in this area. An ectopic ureterocele that is not removed or only partly excised in the region of the vesical neck may act as a valve hindering normal micturition with all the obvious consequences.

In our experience with organ-preserving operations, the best results were produced by operative interventions in which the ureterocele was excised together with the involved ureter and ureteroneopyelostomy or uretero-ureterostomy was performed. An antireflux operation is required in such cases to prevent VUR into the adjacent ureter. These operations remove the cause of the disease

and provide for good flow of urine through the adjacent ureter. Cases discussed in the literature support our impressions of these operations (Stefan, 1966).

After surgery for ureterocele, all patients receive long-term antibacterial therapy until repeated tests show that the urine contains no abnormal admixtures. The remote results are studied by means of excretory urography, cystoscopy, and cystourethrography.

Retrocaval ureter is a rather rare malposition of the right ureter associated with a developmental anomaly of the vena cava inferior. During the developmental process three pairs of primary veins arising in succession (vv. cardinales, vv. subcardinales, and vv. supracardinales) form the trunk of the vena cava inferior, the suprarenal segment developing apart from the postrenal segment (Maksimenko, 1949). In the initial period of embryonal development the venous system is symmetrical but later it becomes asymmetrical. The left primary veins of the vena cava inferior system reduce and some transform into the veins of the kidney, suprarenal gland and testis. This complex process is linked with the involution of the mesonephros and the formation of the permanent kidney. The three pairs of successively arising primary veins richly anastomosing with each other contribute to the development of connections which are important in the origin of some pathological conditions. The processes of reduction and transformation occur mainly on the left (Maksimenko, 1949).

The kidney and ureter are at first surrounded by a venous system, but then ascending and moving laterally, they leave the venous ring formed by the anterior and posterior branches of the above-named veins.

If the right subcardinal vein does not reduce, it develops into the vena cava inferior which thus compresses the ureter. This transformation and retrocaval position of the ureter are represented schematically in Fig. 122.

The clinical manifestations of a retrocaval ureter are linked with its compression by the vena cava inferior, which results in disorders of the flow of urine and the development of hydronephrotic transformation, pyelonephritis, and hypertension and the formation of calculi. The patients' complaints are the same as those encountered in these diseases. The diagnosis is based on the findings of excretory urography and retrograde ureteropyelography. The exact diagnosis is made, however, when venocavagraphy is undertaken in addition to these methods of examination. The shadow of the retrocaval ureter in the upper third is usually superimposed on the shadows of the lumbar vertebrae. In other words, the ureter distal to the pyeloureteral segment is displaced

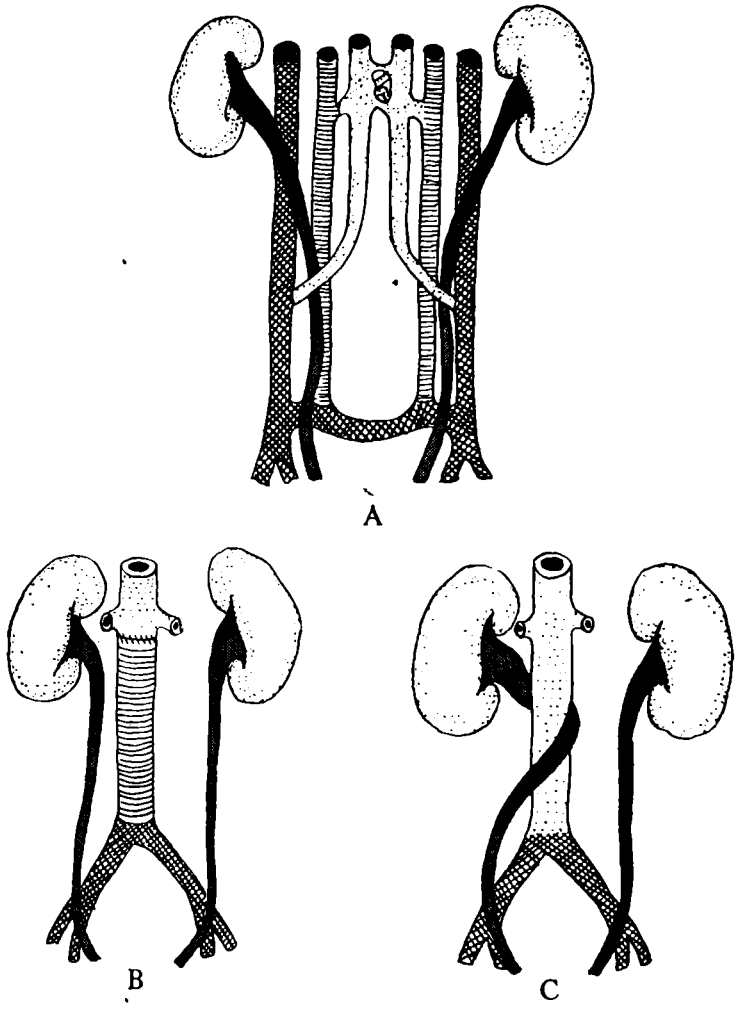


Fig. 122. Development of the vena cava inferior represented schematically after Hollinshead (1971).
A, embryonal period; *B*, normal development of vena cava inferior from the subcardinal veins, which is the cause of a retrocaval ureter, *(C)*

medially and then, forming an arc (Fig. 123), it again stretches laterally. As a result, in the middle third the ureter resembles the letter S. The upper part of this curvature widens and is continuous with a drastically dilated, vertically positioned and elongated renal pelvis. We believe excretory urography to be an obligatory procedure for examining the function and the anatomo-morphological condition of a kidney with a retrocaval ureter, as well as the contralateral kidney (Fig. 124).

The anomaly is treated only surgically. Judging from the data in the literature, an organ-removing operation is rarely applied. Reconstructive operations are performed in most cases: resection of the vena cava inferior, freeing the ureter from under it, and

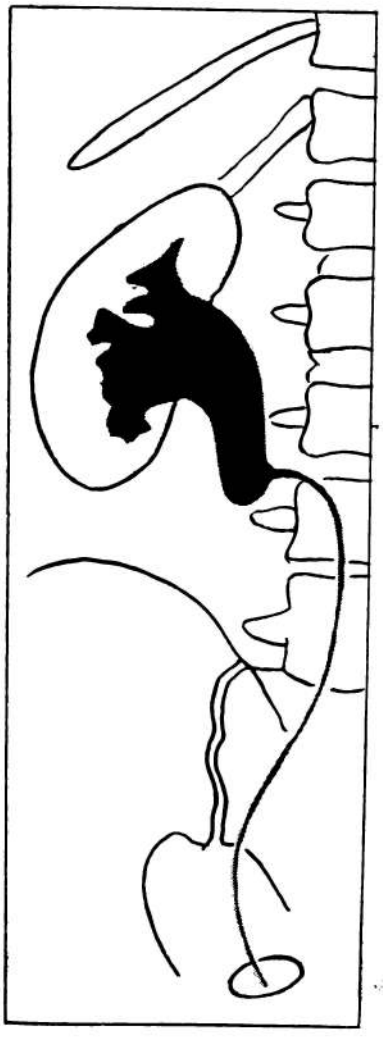


Fig. 123. Retrograde ureteropyelogram and its schematic representation. Retrocaval ureter discussed in the text

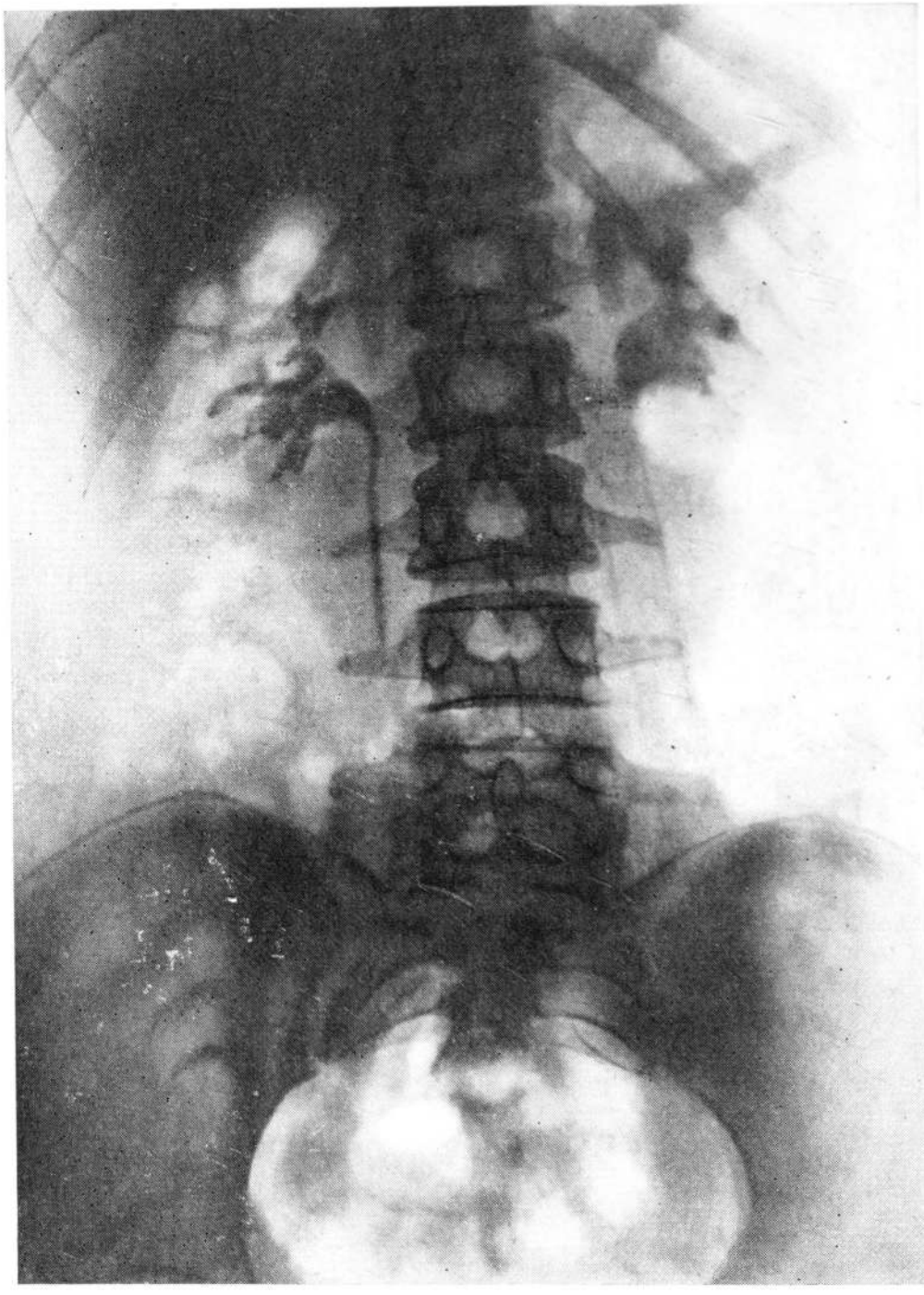


Fig. 124. Excretory urogram. Retrocaval ureter. Dilatated pyelo-calyceal system on the right. The renal pelvis and part of the ureter are dilated and outlined down to the level of the lower border of the third lumbar vertebra. The function of the left kidney is satisfactory

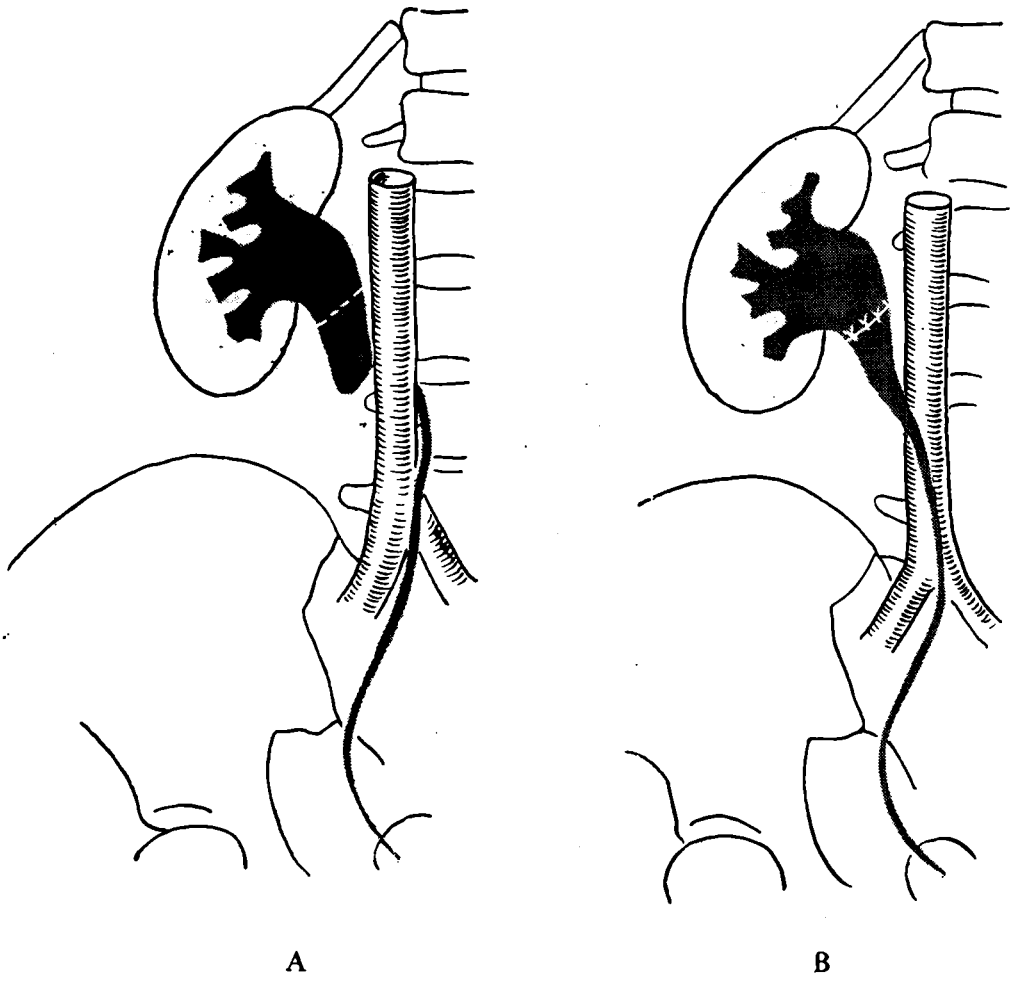


Fig. 125. Diagram of operation for pyelo-pyelostomy in retrocaval ureter. *A*, before operation; *B*, after operation

re-anastomosis of the vein; resection of the ureter with subsequent uretero-ureterostomy in front of the vena cava; resection of the ureter with subsequent ureterocystoneostomy; resection of the renal pelvis with subsequent pyelo-pyelostomy. It should be mentioned here that there is no need for resection of the vena cava inferior as well as ureterocystoneostomy in cases of retrocaval ureter. The most expedient operations are resection of the ureter and subsequent uretero-ureterostomy (Kan, 1963) which we performed in one case, and resection of the pelvis with subsequent pyelo-pyelostomy by means of which we treated two patients. We prefer the last operation because there is no danger of ureteral stricture and, besides, in cases of infection of the urine (Fig. 125) and stones in the kidney the renal pelvis can be drained without conducting another operation. In one of our cases the pelvis had to be drained.

Herniation of the ureter. In this abnormality in the course of the ureter, the ureter is a component of an inguinal, inguinoscrotal or femoral hernia. Dourmashkin (1937) collected the most data from the literature on ureteral herniation; he notes that in this anomaly the ureter is elongated and part of the elongated and convoluted ureter is found in the hernial sac.

The symptoms of ureteral herniation are linked with disturbed flow of urine and concomitant infection: pain, pyuria, fever during the exacerbation of inflammation and strangulation of the hernial contents. The diagnosis is made by means of excretory urography and retrograde pyelography, which show the image of an elongated and tortuous ureter which is displaced downward. If the function of the kidney is preserved, resection of the excess length of the ureter and uretero-ureterostomy or ureterocystoneostomy are indicated. Nephro-ureterectomy is carried out in cases of serious anatomo-functional changes.

Anomalies in the calibre, shape, and form of ureters. These anomalies form a group of various rarely encountered ureteral malformations. They include aplasia and hypoplasia of the ureters, congenital strictures, valves and diverticula, spiral ureter, and neuromuscular dysplasia of the ureter (achalasia, megaloureter). Some authors also include in this group impaired patency of the pyeloureteral segment manifested in early childhood (congenital hydronephrosis); the possibility of this disorder being consequent on an inflammatory process (strands constricting the pyeloureteral segment) can hardly be ruled out. Our experience shows that vesicorenal refluxes occurring in early childhood are often conducive to the development of secondary changes in different parts of the ureter, in the pyeloureteral segment among others.

Aplasia or hypoplasia of the ureter is accompanied by renal aplasia or hypoplasia. In such cases the ureter is a thin fibrous strand or is very thin. The trigone of the bladder is usually also changed, while the ureteral orifices are narrow or have not formed at all and are seen as blind depressions. Even a No. 2 ureteral catheter (Charrièr's scale) often cannot be introduced into the ureter. Excretory and retrograde ureteropyelography help in the recognition of this developmental anomaly; they demonstrate a very narrow ureteral lumen in hypoplasia or no lumen in aplasia. In hypoplasia it is advisable to introduce the opposite end of the catheter into the ureteral orifice so that the contrast medium can flow easily into the lumen of the thinned ureter. Otherwise the wall of the thin ureter will cover the lateral opening in the ureteral catheter and the contrast medium will not pass up along the ureter. We use this method also when there are obturating calculi in the intramural segment of the ureter and in ureteral

strictures. In some cases renal arteriography is the main diagnostic method.

Usually no treatment is required for ureteral aplasia. Hydronephrosis, renal atrophy, and hypertension may develop in hypoplasia and nephro-ureterectomy or ordinary nephrectomy then has to be carried out. Pytel and Chebanyuk (1969) also point to the possible formation of a cyst in the occluded segment of the ureter.

Congenital ureteral stricture. This anomaly is usually characterized by fibrosis of the ureteral urothelium on a limited area without involvement of the muscular and adventitious coats. Congenital stricture of the ureter is mostly unilateral and solitary but bilateral and multiple strictures are encountered. The vesico-ureteral and pyelo-ureteral segments are affected most frequently. It prevails among boys.

It is general knowledge that as the kidney ascends and rotates narrowed areas form in the ureter, which are considered physiological constrictions. These are the pyelo-ureteral segment, the site where the ureter crosses the iliac vessels, and the intramural part and orifice of the ureter. These constrictions are of course not identical with cystoid-segments. Constrictions in other parts of the ureter should therefore be regarded as developmental anomalies characterized by deficient width of their lumen. The mucosa in the region of the physiological constrictions is normal and there is no stasis of urine in the upper urinary tract proximal to them; in congenital ureteral stricture, in contrast, mucosal folds or thickenings are found for a length of 1 to 3 cm and there is urinary stasis in the proximal parts of the upper urinary tract. Depending on the degree and level of the constriction, congenital strictures of the ureter are attended by hydronephrosis or hydroureteronephrosis.

The clinical manifestations of the anomaly are the same as those of the diseases and complications mentioned above. But since this developmental ureteral anomaly is congenital and manifested in early childhood, its clinical picture is very diverse. Patients are troubled by abdominal pain, gastro-intestinal disorders, dysuria, lumbar pain, elevation of temperature, etc. Infants who have neither dysuria nor abnormalities in the urine are treated by paediatricians for a variety of erroneously diagnosed diseases. Abnormal urine values and localized lumbar pain suggest a disease of the kidneys and urinary tract.

The diagnosis is made by means of excretory urography and retrograde ureteropyelography. The treatment of congenital ureteral stricture is mainly surgical and depends on the condition of the kidneys and urinary tract and on the level of the obstruction. In the initial stages of the disease, when kidney function

is maintained, the ureter is catheterized, anti-inflammatory treatment is conducted, and resolvents are prescribed.

Congenital ureteral valve. Östling (1942) contends that there are actually more people with valve-like folds of the ureteral mucosa than is recognized; these folds are found proximal to the vesicoureteral segment but rarely disturb the passage of urine. Only a few reports of congenital ureteral valves have been published. Ioffe (1961) collected ten cases from the world literature. The literature sources show that the ureteral valve is a transversely lying mucosal fold containing muscle fibres. By the fourth month of life these folds usually resolve, but if this does not occur a valve remains. A circular mucosal fold with villi is then discovered in the ureteral lumen. Histological examination of this area reveals thickening of the muscular coat and, on the edges, loose fibrous tissue and preserved transitional epithelium.

The symptoms of the disease are similar to those of hydronephrosis, hydroureteronephrosis and chronic pyelonephritis. The diagnosis is based on the discovery of a transverse defect during excretory urography (if the function of the kidney is preserved) or, more frequently, during ascending ureteropyelography. Similar functional and anatomical changes are also found in the upper urinary tract in other pathological conditions which is why a final diagnosis is made on the basis of the results of the histological examination of specimens obtained during surgery.

Treatment consists of the resection of the ureteral segment with the valve and creation of end-to-end or side-to-side ureteral anastomosis. Ureterocystoneostomy or Boari's operation is performed in valves of the lower parts of the ureter. Nephro-ureterectomy is carried out if the kidney does not function.

Congenital ureteral diverticulum. This condition is marked by a cyst-like distension of the ureteral wall. It is mostly encountered in the lower third of the ureter and its origin is linked with the formation of an accessory ureteral bud. Sometimes the diverticulum has to be differentiated from splitting of the ureter with a blindly terminating branch in the proximal segment. Congenital and acquired diverticula are distinguished; the disease may be bilateral. The wall of the diverticulum has a muscular and a mucous coat. A ureteral diverticulum causes occlusion of the urinary tract proximal to it and consequent hydronephrosis. The clinical picture is that of hydronephrosis or hydro-ureteronephrosis with or without infection of the urinary tract. The formation of calculi or sedimentation of salts in the diverticulum is also possible. The diagnosis is made from the findings of excretory urography, although retrograde ureteropyelography is more demonstrative.

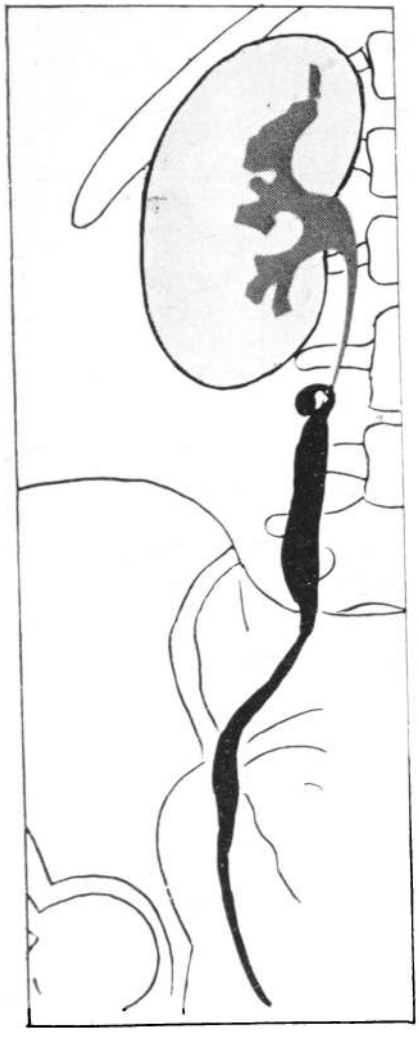


Fig. 126. Retrograde ureteropyelogram and its schematic representation. Spiral ureter. Operation for ureterolysis

Treatment comprises the resection of the diverticulum and uretero-ureterostomy or ureterocystoneostomy. Nephro-ureterectomy is necessary if the kidney does not function.

Spiral ureter. The formation of a spiral ureter is attributed to the fact that only the kidney, without the ureter, rotates in the embryonal period. This developmental anomaly can be unilateral or bilateral. The ureter can have a spiral structure with varicosity of the veins in the retroperitoneal space, or of the seminal or ovarian veins. Restricted ureteral torsion may be encountered, producing what is called a circular ureter (Pytel and Chebanyuk, 1969). The twisted ureter creates an obstacle for normal urine drainage. We had one female patient with a twisted ureter, she complained of dull lumbar pain which became intense at times. The elimination of indigo carmine and contrast medium is delayed and sometimes these methods cannot reveal the developmental anomaly. Retrograde ureteropyelography is the main diagnostic method (Fig. 126).

Treatment is surgical and depends on the character of the ureteral structure, the length and the condition of the upper urinary tract.

Neuromuscular ureteral dysplasia is a combination of congenital stricture of the orifice and intramural part of the ureter with neuromuscular dysplasia of the distal ureteral cystoid. These factors, as well as disorders of ureteral tonus and dynamics, cause elongation and dilatation of the ureter. Dilatation of the lower ureteral cystoid is known as achalasia; dilatation of the whole ureter is called megaloureter. Dilatation of the ureter in cases of abnormalities of the neck of the bladder or the urethra with vesico-ureteral reflux should be considered secondary. It is difficult to distinguish them, particularly if there is concomitant infection.

The diagnosis of neuromuscular dysplasia of the ureter is difficult because there are no complaints in many cases. Patients are given drugs for many years because of vague lumbar or abdominal pain, dysuria, and pyuria. The developing symptoms of renal insufficiency are interpreted as manifestations of pyelonephritis. Only timely urological examination helps in revealing the true disease.

The results of urological examination allow the diagnosis of a giant ureter to be made quite easily. The examination includes cystoscopy, cystography, and retrograde or excretory (descending) urography. Cystoscopy reveals hyperaemia of the cystic mucosa and gaping (sometimes widely) ureteral orifices. Indigo carmine flows from the orifices of the ureters in a sluggish stream and stains the urine weakly. If the kidney functions an excretory urogram shows a clear image of the tortuous and drastically

dilated ureter with a relatively mild dilatation of the pelvis. Depending on the function of the kidney, the picture is clear or obscured (Figs. 127 and 128). Urotelescopy and cineurography together with cystography help in detecting the vesico-ureteral reflux. In a non-functioning kidney all these changes are demonstrated by retrograde pyelography.

Megaloureter must be treated surgically. The degree of surgical intervention is determined to a great extent by the shape of the megaloureter, the duration of the disease, and the severity of the pathological process. The disease is often found in both kidneys, which is why an attempt should be made to carry out organ-preserving operations. The outcomes of treatment are best in the first and second stages of the disease.

Many different methods have been suggested for correcting megaloureter. Most urologists recommend resection of the juxtavesical segment of the ureter and its reimplantation into the bladder (Lopatkin and Derevyanko, 1969). Nephro-ureterectomy has to be carried out when the kidney does not function and has suffered irreparable morphological changes.

During surgery it is necessary to create conditions for the normalization of urodynamics and correction of vesico-ureteral reflux. This is accomplished by reimplantation of the ureter into the urinary bladder by means of direct or indirect ureterocystoneostomy (Boari's or Demel's operation). In direct ureterocystoneostomy a graft of the cystic wall is used for forming a semi-cuff or a sphincter-cuff on the transplanted ureter. In indirect ureterocystoneostomy the ureter is passed under the mucous membrane or under the muscle so as to prevent a reflux.

Pugachev (1977) claims that the principal objects of reconstructive plastic operations on a megaloureter are as follows: (1) resection of the affected ureteral segment creating the obstacle; (2) modelling of the ureteral diameter to normal size; (3) neoimplantation of the ureter into the bladder and performance of an antireflux operation. One must agree that VUR with all its consequences is a frequent complication after surgery on a megaloureter if an antireflux operation is not carried out, whereas antireflux operations prove successful provided that the ureter has a normal diameter and is not strangulated by the cystic wall during the operation.

Judging from the reports in the literature, Bischoff's operation (1961) has gained wide popularity in the management of megaloureter. It is indicated when there is an obstacle in the intramural or juxtavesical parts of the ureter. The distal segment of the dilated ureter is excised and ureteroneocystostomy is performed. Today many surgeons use the Ledbetter-Politano antireflux technique in this operation. The operation is performed

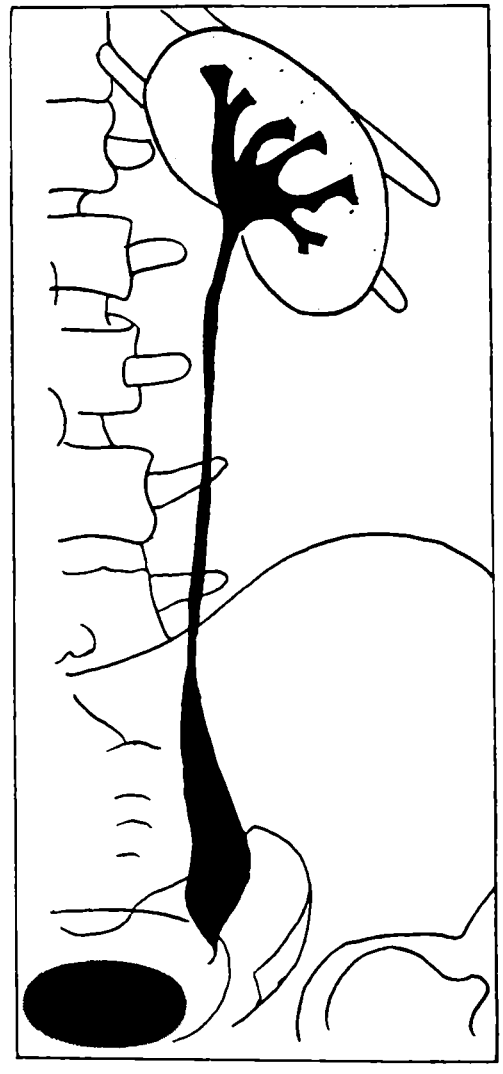
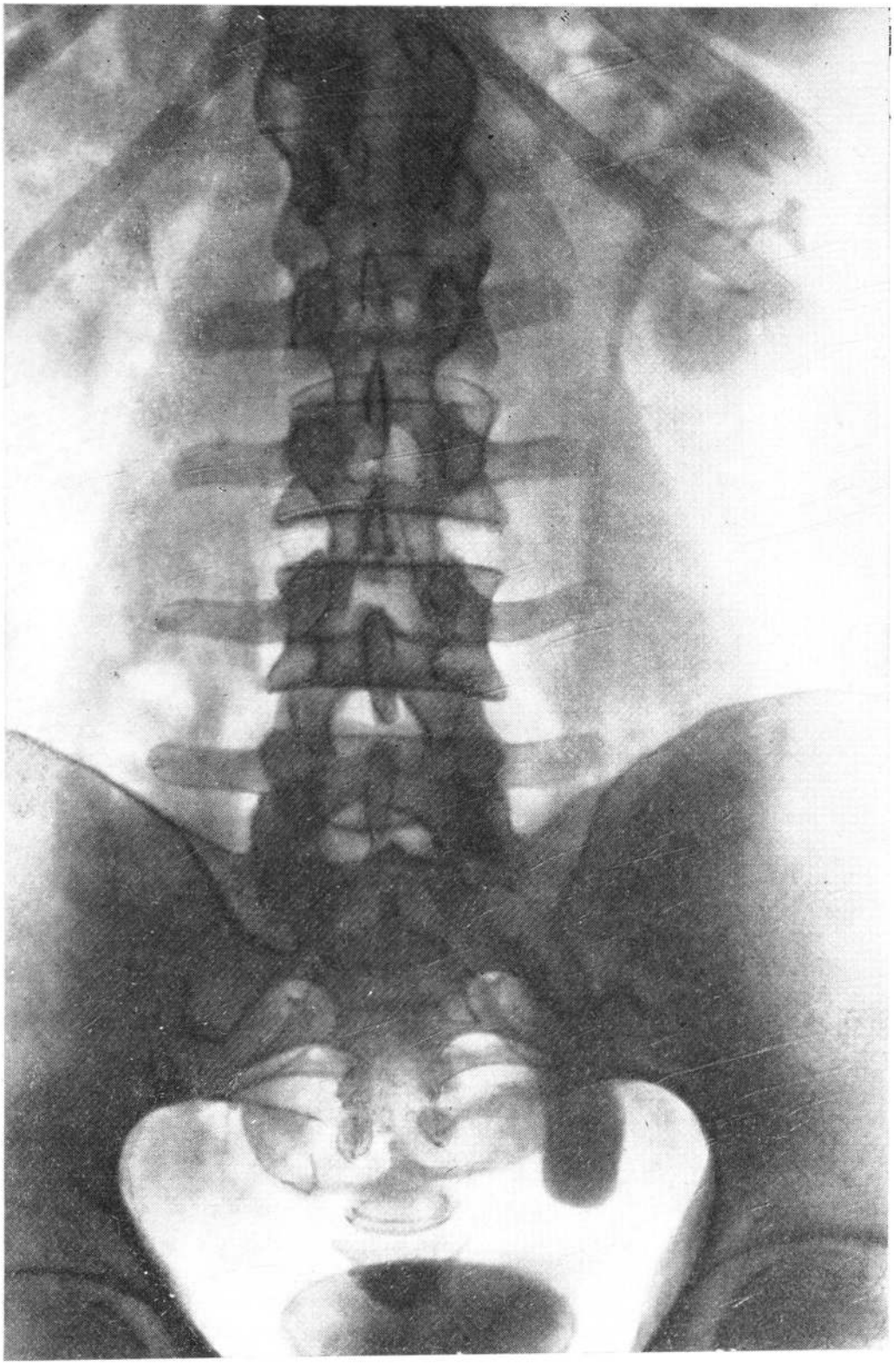


Fig. 127. Excretory urogram. Ureteral achalasia on the left, only the lower cystoid is involved ▶

extravesically or transvesically. It is our experience that this technique produces the best results.

Gregoir's (1962) method is also used in the management of this disease. The ureteral segment, mobilized beforehand, is sunk into the wall of the bladder. The terminal segment of the ureter is separated from the posterior wall of the bladder. An incision is made in the wall through the detrusor and down to the mucous coat from the site where the ureter enters the bladder and in the direction of the apex for a distance of 3 cm. The ureteral segment is then fitted into the canal thus formed and the wall of the bladder is sutured over it. This operation is advisable in cases of



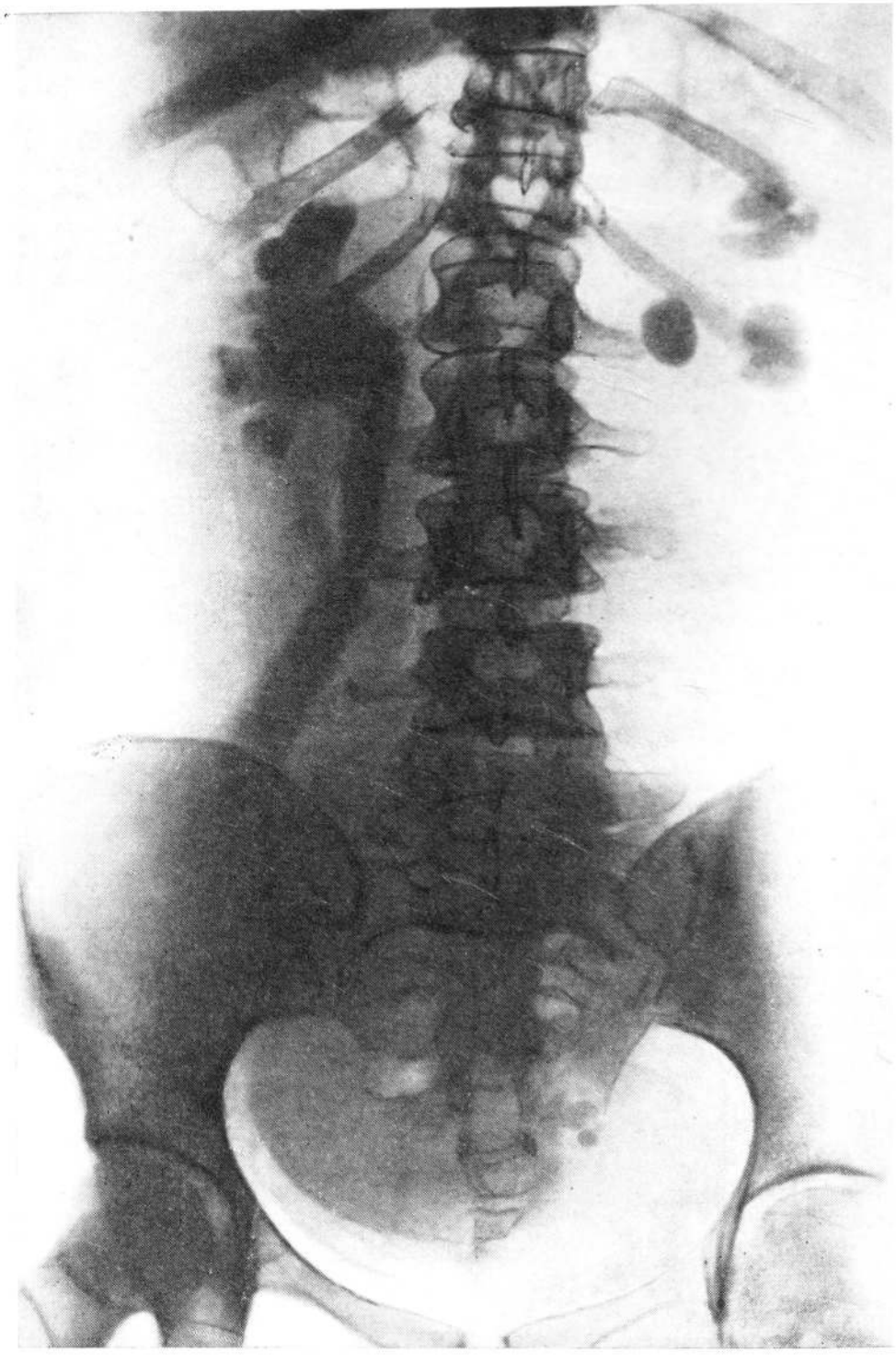


Fig. 128. Excretory urogram. Achalasia of both ureters. Sharply diminished function of the left kidney; shadows of only two calyces and a migrating calculus in the renal pelvis are demonstrated

mild dilatation of the ureter. In severe morphological changes in the ureteral segment VUR may develop after it.

When the ureter is greatly dilated and its contractility is considerably changed, it is advisable first to relieve the urinary tract from its load by means of ureterocutaneostomy or nephrostomy. If the anatomo-functional changes are compensated in this way, an organ-preserving operation may then be carried out during which part of the ureter is removed by one of the methods discussed and the ureter is implanted again into the urinary bladder with antireflux protection. With this two-stage surgical intervention, the kidney is preserved even in very difficult cases. It yields quite good remote results.

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